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Leber congenital amaurosis - a case report

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Purpose To present a rare, inherited retinal dystrophy with early-onset severe visual impairment.

Methods A 13 month-old girl with lack of visual attention since 3-4 months of age attended our department. Her pupil reactions were sluggish and she had slight roving eye movements. Examination methods included indirect binocular funduscopy, measuring refraction and electrophysiology. Follow-up: 12 month after the first examination.

Results At the age of 13 months the only retinal alteration was a marbleized pattern at the periphery. At the age of 18 months funduscopy revealed diffuse, confluent whitish spots except in the central area within the vascular arcades. Full-field electroretinography was not detectable under either scotopic or photopic conditions at repeated examinations.

Conclusion The reported case was clinically diagnosed as leber congenital amaurosis on the basis of ERG-examinations. In cases of unexplained, severe visual impairment in preverbal infants, electrophysiological tests are of prime importance.

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Five-year mortality rate following blind/partially sighted registration in Sunderland, UK

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Purpose To compare five-year mortality rate of the patients registered blind/partially sighted (BD8) with mortality in the general population.

Methods Retrospective study of patients registered blind/partially sighted during one-year period in a UK eye hospital. The data was collected from the hospital records and the death registry for the area covered by the hospital. The age at BD8 registration and death, the cause of BD8 and sex were recorded. The data was age and sex stratified, according to the UK national statistics office for calculating death rates. Statistical analysis was done with and without patients with diabetes and cardiovascular events, using chi-squared test.

Results 153 patients were BD8 registered in 1995 by the hospital: 92 (60.13%) females and 61 (39.87%) males. The mean age at registration was 73.37 years for the whole sample. However, the mean age at BD8 registration for males was 67.04 years and 77.56 years for females. The mean age at death was 82.55 years for the whole sample and 77.7 years for males and 85.77 years for females. The number of deaths in each age group was evenly distributed for the males; the female deaths were concentrated in the older age group.

Conclusion The expected death rates in our sample were lower than expected compared with the general population for both female/male subgroup and the whole group. It was also observed in this sample that the younger at BD8 registration (less than 65 years of age) the more likely to die if visually impaired than general population. This might be because the patients registered blind/partially sighted younger had a higher incidence of cardiovascular disease and diabetes.

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The Prevalence of Refractive Errors in University Students in Portugal

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Purpose This study was initiated to assess the prevalence of refractive state in university student's population in Portugal.

Methods A population sample of 199 university students were randomly selected and examined in cycloplegia. The results obtained using the subjective distance refractive method were used in the analysis and refractive values were converted into spherical equivalents (SER) for some analysis (i.e. sphere value + 1/2 of the cylinder value). Myopia was defined as SER < -0.25 D, emmetropia as SER >= -0.25 D and <= +0.25 D and hyperopia as SER > +0.25 D.

Results The refractive error of the sample, ranged from -8.75 to +3.75 D mean spherical equivalent + 0.20 ± 1.53D (Mean ± SD). The maximum amount of astigmatism was -2.25 D. The incidence of refractive errors was: 22.1% of the students had myopia, 23.1% had emmetropia and 54.8% had hyperopia. Statistically there are no significant differences between male and female (male mean refractive error (MRE) = 0.21 ± 1.55 D; female MRE = -0.06 ± 1.59 D) or any significant MRE differences between the students in the first year or in the last year (1st year (MRE) = 0.051 ± 1.685 D; 4th year MRE = -0.03 ± 1.29 D). However, the prevalence rates of myopia in females (26%) are higher than the rates in males (14%). And the prevalence rates of hyperopia in 1st year students (57%) are higher than the rates in the 4th year (49%).

Conclusion The results then show an incidence of myopia similar to results obtained in other countries. Just like in other studies, ours also revealed a higher prevalence of myopia among women than men, as well as show a myopia shift during the university course.

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The prevalence of open angle glaucoma on the west coast of Iceland

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Purpose To establish the age- and sex-specific prevalence of open-angle glaucoma (OAG) on the west coast of Iceland.

Methods The compulsory ophthalmologic examination needed for prescription of eye glasses in combination with information obtained from Statistics Iceland were used to retrospectively establish the glaucoma prevalence in the town of Akranes and to estimate the lowest prevalence for the greater west coast area.

Results 79.1% of 1443 inhabitants > 50 years of age in Akranes had visited the eye clinic at least once during the past 5 years. The overall prevalence of OAG in this population was 4.8% (95% CI 3.6-6.1) with no significant difference between sexes. The lowest prevalence calculated for the west coast as a whole was 3.8% (95% CI 3.2-4.4). The prevalence of OAG increases with increasing age (p<0.001). The predominant glaucoma types were simple and capsular glaucoma, similarly distributed.

Conclusion The results with respect to glaucoma prevalence and participation rate are in the same magnitude as in similar studies indicating that the method used in this study is comparable to other methods used to establish glaucoma prevalence.