Strabismus, Susceptibility to

Alternative Names
Strabismus, Susceptibility to, 1
STBMS1

WHO International Classification of Diseases
Diseases of the eye and adnexa

OMIM Number
185100

Mode of Inheritance
Familial, but no simple Mendelian inheritance established

Gene Map Locus
7p22.1

Description
Strabismus, commonly known as ‘squint’ or ‘cross-eye’ is a misalignment of the visual axes of the eye, usually caused by a lack of coordination between the extraocular muscles. As a result, the eyes look in different directions, and cannot focus simultaneously on a single point, preventing binocular vision, and affecting depth perception. Strabismus may be of different types. In incomitant strabismus, the degree of ocular misalignment differs with the direction of gaze; whereas in concomitant strabismus, the degree remains the same, irrespective of the direction or the eye which is fixing. Amblyopia is a complication of strabismus, in which the brain learns to ignore the messages from one of the eyes, leading to loss of vision in that eye. Strabismus may appear in an individual by itself, or as a part of one of several syndromes, such as Duane’s syndrome, Brown’s syndrome, Mobius syndrome, and sixth nerve palsy.

Various studies in different populations have shown a prevalence of 1-5% for strabismus in the general population. A comprehensive eye examination can easily detect strabismus. Some special tests, such as prism test and Krimsky test are also used to evaluate the degree of alignment of the eyes. Correction of the abnormality requires ocular exercises to strengthen the muscles. Glasses may also be prescribed. In case of a failure to correct the misalignment by exercise alone, surgery can be performed. Amblyopia is corrected by patching the preferred eye, so that the weak eye is forced to be used.

Molecular Genetics
Several studies have conclusively shown the genetic etiology of strabismus, with an average of 37% of the cases showing a positive family history. Linkage of the condition to gene locus 7p22.2 was shown in a large family with strabismus. However, the disorder shows genetic heterogeneity, with chances of more than one gene being involved in its development.

Epidemiology in the Arab World

Egypt
Shawky et al. (1990) studied the role of heredity, prenatal, and parental factors in the development of strabismus in 42 patients (male:female ratio 0.8:1) with primary concomitant strabismus, and four patients with strabismus as part of a syndrome. The patients were subjected to full history taking, thorough clinical examination, condition of paternal eye movement, pedigree construction, and segregation and consanguinity analysis. As a control group, 35 age and sex matched cases, without strabismus, were also examined. Segregation analysis involved testing for autosomal dominant inheritance using Neel and Schull’s hypothesis, autosomal recessive inheritance using Fisher’s equation, X-linked inheritance by pedigree inspection, and Emery’s criteria for multifactorial inheritance. The cases of convergent strabismus were found to be significantly higher than divergent strabismus.
Hypermegopia was found to be the most common error of refraction (59.5%), followed by hypermetropia with astigmatism (21.4%), and myopia, (14.3%). No significant relationship was found between occurrence of strabismus and drug consumption during pregnancy, maternal age, or paternal age. On the other hand, a significant association was seen between the patients and occurrence of latent or manifest strabismus in the mothers (33.3% mothers showing latent and 13.3% showing manifest strabismus). Shawky et al. (1990) found that affected mothers with latent or manifest strabismus had a four times more risk to have affected children as compared to normal mothers. Only eight children, out of the 63 offspring of affected parents, were similarly affected, ruling out autosomal dominant inheritance. Similarly, application of Fisher’s equation ruled out the autosomal recessive mode of inheritance too. The appearance of affected males with non-affected daughters, and at least one case of male to male transmission was inconsistent with an X-linked mode of inheritance. The sharp fall in the frequency of affected relatives from first degree to second degree to third degree relatives in the pedigree analysis, higher frequency of affected individuals among relatives of severely affected cases, a shift to the right for the hypothetical curve of liability, all were found to point out to the multifactorial mode of inheritance for the condition. Heritability was estimated to be 41.4% among first degree relatives by Shawky et al. (1990).

Oman
Lithander (1998) conducted a nationwide school survey to investigate the prevalence of common eye disorders in the Sultanate of Oman; mainly amblyopia induced by strabismus or anisometropia. In a sample that included 49 schools with 6541 children in Grades 1 and 6, examinations of the eyes were conducted in 96% of children using visual acuity screening to 0.5. All 6292 children had an external eye examination. All children failing the visual acuity test had a complete eye examination. Amblyopia was found in 0.92%, strabismic amblyopia in 0.48%, anisometropic amblyopia in 0.44% and strabismus with or without amblyopia was seen in 0.9%. There was statistically significantly (p<0.05) more amblyopia in 11-12 year olds (Grade 6) when compared with the 6-7 year olds (Grade 1). This difference was only found among the strabismic children.

In 2004, Khandekar and Abdu-Helmi reviewed school screening and refraction data to estimate the magnitude and determinants of refractive error in school children. Trained physicians screened 416,157 students to evaluate their visual status and identified 28,765 students with defective vision. Refractionists refracted 25,733 (89.5%) of them, determined the refractive error and prescribed spectacles. Students with ocular co-morbidity and visual disability were re-examined and treated by the ophthalmologists. The prevalence of myopia was 4.1% (95% confidence interval [CI] 4.06-4.18). It was higher among female than male students [rate ratio (RR) 1.69 (95% CI 1.64-1.74)]. The rate was more in students of higher age groups (chi2 = 11,179 degrees of freedom = 2 p<0.00001). Regional variation in myopic trend was marked. The prevalence of hypermetropia was 0.4% (95% CI 0.37-0.41). However, it could be an underestimation as presence of accommodative spasm was not taken into account. The risk of low vision disability was significantly higher in male students than female students. The prevalence of amblyopia was 0.3%. It was significantly higher in male than female students. First primary students had strabismus of 0.5%.

References

Contributors
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