

Refractive Errors and Visual Anomalies

As Related To The Degree of Retardation In A Down's Syndrome Population

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Abstract

A group of 86 children diagnosed as having Down's Syndrome were evaluated as to their refractive and visual status. The results of these evaluations were compared with previously obtained data as to the level of retardation of each subject. In the areas of ocular pathology and strabismus, there was a high statistical correlation (p.01) between the prevalence of these conditions and the level of retardation. Refractive errors did not follow this pattern. Possible reasons for this are discussed along with the general status of and need for visual evaluations in this population.

Key words:

Down's syndrome, refractive error, intelligence, strabismus, pathology

There have been numerous studies examining the frequency of various visual defects in mentally-retarded individuals in general and in the Down's Syndrome population in particular.¹⁻⁹ Some of these have focused mainly on the frequency of various pathologic conditions such as cataracts, keratoconus, and iris defects, while others have examined the area of strabismus and refractive error. Research reports have also appeared detailing the effects visual defects have on the functional abilities of the retarded.¹⁰ There have also been some studies with a wider scope that included all of the above areas of concern. However, as far as I have been able to determine, there has been no attempt to relate these defects to the level of retardation, possibly because there was no reason to presuppose that such a link could or should exist. This study was specifically designed to take this factor into consideration and to examine the possibility of such a connection.

Visual defects in the mentally retarded population as a whole and the Down's Syndrome population within it, frequently go unreported or at best under-reported. Most studies in this area conclude that one of the most striking features are undiagnosed vision disorders.^{4,7} One reason for this is that it can be difficult for parents to find a vision care provider who is willing to make the necessary effort to evaluate their child.⁴ An additional barrier to appropriate diagnosis and treatment can be

the belief that this is probably the "least of the child's problems" and certainly not an area of major import. This attitude ignores the possible crucial role that the child's vision can play in his intellectual development and the importance of making the care givers/educators aware of any limits on the child's or adult's visual system.⁹⁻¹⁴

Subjects

The group of Down's Syndrome children included in this report were part of a larger (approximately 1,000 subjects) group of retarded children whose findings will be reported in a subsequent study. As this group was easily separated out of the general group and had an established prevalence of visual anomalies, it was felt that it would be easiest to initially study this group.

All the subjects in this study had been tested previously for cause of retardation and quantified as to level of intelligence. Thus, this sample had already been diagnosed as to retardation syndrome and level of intelligence but virtually never had received an optometric or ophthalmologic evaluation. The children were grouped into four categories:

- * Mildly retarded: educable (Level 1)
- * Moderately retarded: trainable (Level 2)
- * Severely retarded: it may be possible to attain ADL (activities for daily living) skills (Level 3)
- * Profoundly retarded: cannot be trained to care for themselves (Level 4)

Over the course of four years, over 1000 patients were evaluated. However, as stated, the current study focuses only on the children in the Down's Syndrome group.

Methods

A total of 86 Down's Syndrome patients were examined at various facilities for the retarded. Distance refractive status was objectively determined either by cycloplegic examination or by using assistants to maximize fixation on a distant target when consent for the use of cycloplegia could not be obtained. Ophthalmoscopy (monocular, direct) was performed along with a cover test (when possible) and external evaluation with a transilluminator. Spectacles were prescribed when applicable and medical referrals were made when necessary. Their ages ranged from 5 years to 18 years with the mean age of 12.5.

The refractive results were divided into three categories:

- * Low myopia/hyperopia (less than or equal to 2.00 diopters)
- * Moderate refractive errors: between 2.25-6.00 diopters
- * High refractive errors: greater than 6.00 diopters.

In the cases of astigmatism, the spherical equivalent was used as it was felt that this method of expressing the uncorrected refractive error would bear the most meaningful relation to the child's performance and intellectual development. For the same reason, in the case of anisometropia, the two eyes were not reported separately, rather, the individual was reported on the basis of the better of the two eyes.

Results

In the case of low refractive errors, as can be seen in Figure 1, there was no clear-cut pattern to the distribution of the refractive error categories. While the mildly retarded group all were in this category, 50% of the moderately retarded, 34% of the severely retarded and 50% of the profoundly retarded also had refractive errors in this range. The moderate refractive errors were almost equally distributed among the moderately, severely and profoundly retarded groups. As can be seen in Figure 2, none of the mildly retarded, 50% of the moderately retarded, 38% of the severe and 42% of the profound were in this category. The one unusual finding ap-

Group	Correlation Coefficient (r)	Significance
Strabismus vs. Pathology	0.988	p.<0.01
Low Ref. Error vs. Pathology	0.818	ns
Mod. Ref. Error vs. Pathology	0.448	ns
High Ref. Error vs. Pathology	0.789	ns

*Using the Pearson r

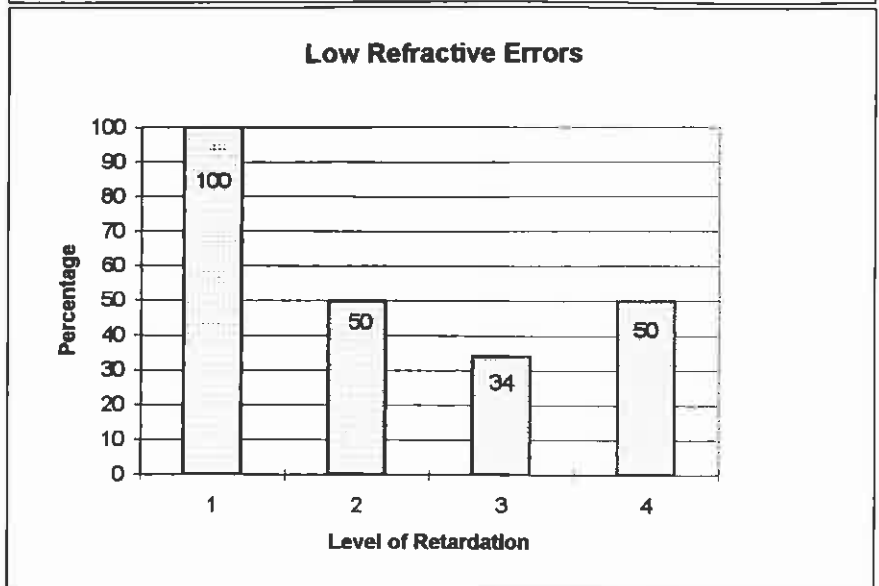


Figure 1.

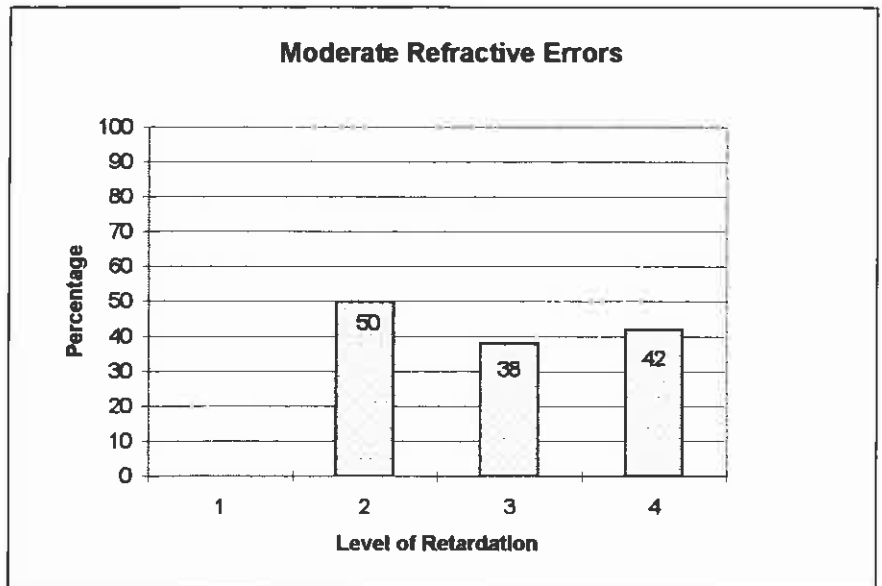


Figure 2.

peared with the high refractive errors which were "over-represented" (28%) in the severely retarded patients (Figure 3). A summary of all these results is presented in Figure 6.

The amounts of strabismus and pathology, on the other hand, seem to follow a very similar and even "logical" course, as is shown in Figures 4 and 5. The more severe the level of intellectual impairment,

the greater the amounts of pathology and strabismus.

In order to obtain a statistical analysis of the data, the prevalence of pathology as related to level of retardation was evaluated first utilizing the Chi Square method. The relationship was significant at the .01 level. Each other category was then compared to the pathology group utilizing the Pearson r method to test correlation. As

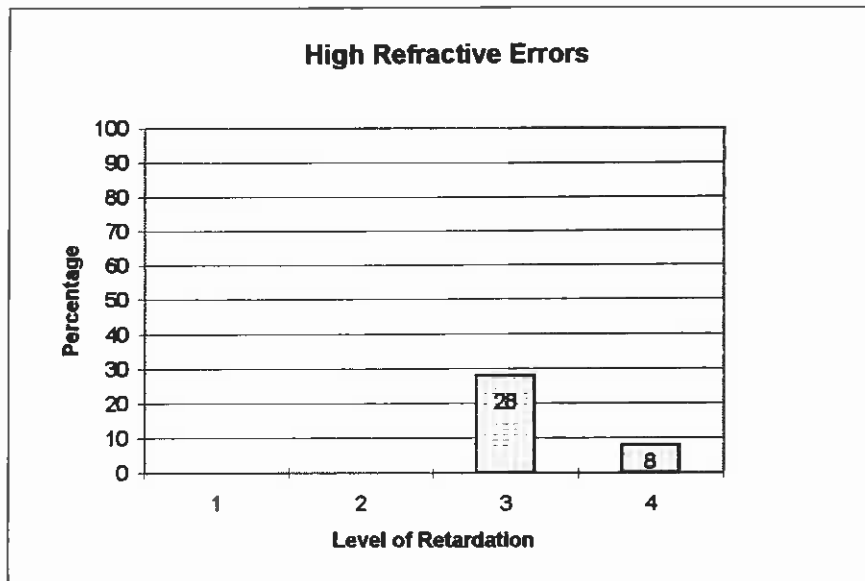


Figure 3.

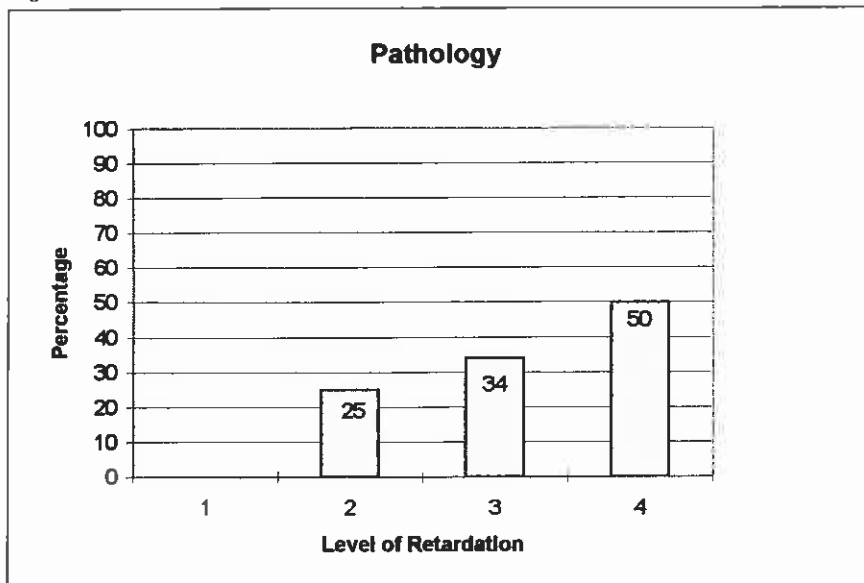


Figure 4.

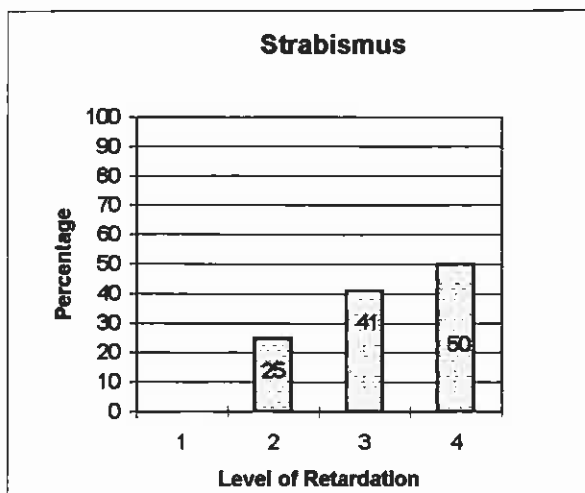


Figure 5.

level $p < .01$ of correlation between the progressive amounts of strabismus and pathology with increasing amounts of mental retardation. However, no such correlation was found in any area of refractive error whether low, moderate or high.

Discussion

The results of amounts of strabismus and pathology seem quite straightforward. There was a continually increasing percentage as a function of degree of retardation. The refractive results follow no such pattern. While low (or no) refractive errors were present

in the entire mildly retarded group, they were present in equal amounts in the moderate and profoundly retarded groups. While moderate refractive errors were absent in the mildly retarded group, they were fairly evenly distributed in the other groups. The one factor that seemed to stand out was the relatively high prevalence of high refractive errors in the severely retarded group (Figure 3).

A possible working hypothesis in predicting the incidence of various anomalies in this population would be to assume that the more severe anomalies would probably be present in those with the greatest level of retardation. In essence that is what was found in the areas of pathology and strabismus. In both instances the amounts rose from 0% in the Level 1 group to 50% in the Level 4 group in graduated steps. This hypothesis was not valid in predicting the distribution of refractive error.

Several hypotheses come to mind to explain the causes for this difference. First it is possible that the factors involving the incidence of pathology and strabismus are more related to each other and to the severity of the syndrome than refractive error. It is also possible that the causes for refractive error are more diffuse than those for strabismus and pathology and, therefore, less inclined to be related to other single factors. However, based on admittedly anecdotal information provided by my work in these settings, i.e., the reaction of the children in the high refractive error group who were provided with spectacles for the first time in their lives, I feel that a different explanation must be considered.

This explanation posits that the uncorrected refractive error can be a major contributing factor in establishing the amount of retardation. This would be especially true if left uncorrected over a long period of time and even more if the amount of the refractive were substantial. One can see this possibility most strikingly in Figure 3 with the "overrepresentation" of high refractive errors in the severely retarded group. It is certainly possible that early correction of these refractive anomalies might have helped move these subjects to the moderate or mild retardation category.

Comments

Whether one accepts the uncorrected refractive error theory or not, one fact stands out in this initial report. A very significant amount of undiagnosed refrac-

tive error, pathology and strabismus existed in this sample. There is no excuse for such lack of care in any subject group and certainly not in one that has known handicaps. The possibility that the data supports the theory that uncorrected significant refractive errors have "docmed" part of this populace to an unnecessarily low level of mental achievement is even more reason to be concerned. It behooves all eye care practitioners to be involved in providing or supporting the provision of care to retarded individuals for as this study and other studies⁴⁻⁷ show, the problems are there.

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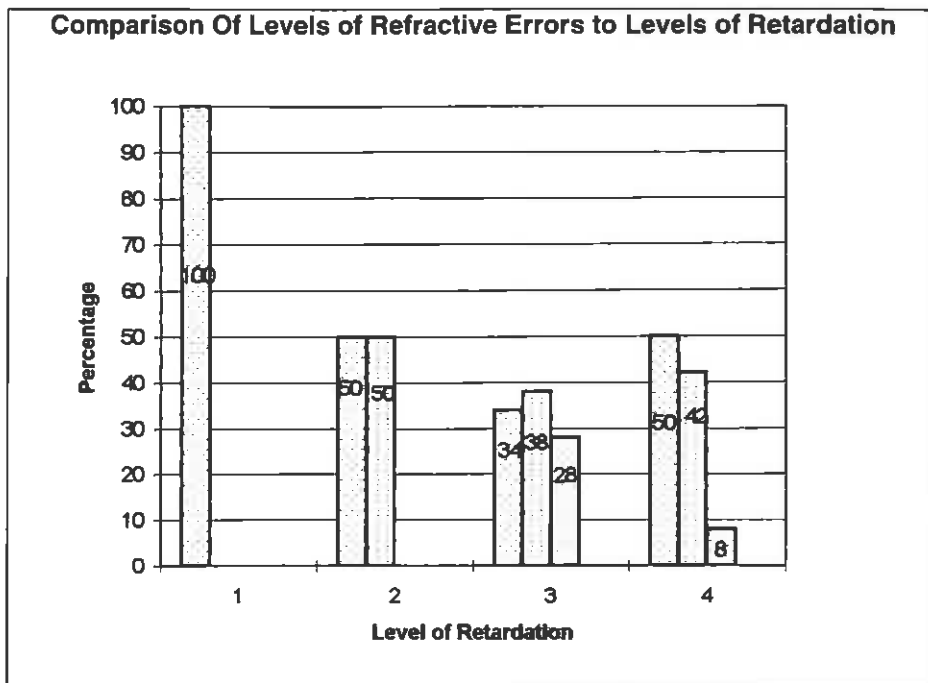


Figure 6.

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