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Epidemiology of strabismus

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In a short paper such as this, it would be a mistake to attempt a comprehensive review of our knowledge of the occurrence of strabismus and those characteristics of populations which may lead to variations in its incidence. To do so would subject us all to mental dyspepsia from a surfeit of information which none of us would have time to consider critically. Instead, I propose to take certain aspects of the epidemiology of strabismus, and to illustrate these by some observations made in Cardiff, with occasional data from other sources.

The sort of questions we can hope to answer by epidemiological studies are those concerned with the prevalence of squint:

How common is it?

Does the prevalence vary from one community to another, and if so, can we identify any characteristics of these communities which may explain the difference?

How do new cases arise, and at what ages?

The first of these questions can be answered reasonably accurately, but the others present difficulties. The common snag in population research is that a study of previously published work reveals that sampling methods and criteria used in definition are either not stated or differ so much from one another that it would be most unwise to draw any conclusions from comparisons of prevalence. As a result, it becomes necessary to use mainly internal comparisons, and in particular the techniques of comparing a group of strabismic patients with a group of non-strabismic patients drawn at random from the original population, and examined by the same techniques. A number of the comparisons I shall present comes into this category.

Material and techniques

A brief description of the nature of our population and our examination procedures and definitions is clearly the first step in describing some of our results.

Our sample consisted of all the children born in the City of Cardiff between January 1 and December 31 of one year. The details of the 4,832 children born within these limits were obtained from the birth records maintained by the Medical Officer of Health, and the children were examined wherever possible at school during their first year by a research orthoptist who carried out a cover test at both distant and near fixation and estimated visual acuity. Those children not available during visits to school were sought out and examined where and when possible, and the coverage thus included private schools and schools for handicapped children.

This technique, with a population largely captive in schools, should be and proved to be capable of giving a very high coverage of the population under study. The final total of children screened was 4,784, almost exactly 99 per cent. of the total sample.

From the results of this initial examination, a selection was made on the basis of cover test results. Those with certain cover test responses were asked to return for a much more detailed orthoptic investigation and the taking of a full history including a number of questions related to the social

rather than to the purely medical background. At the same time, a random sample of children drawn from the remainder of the population was subjected to a similar orthoptic investigation and interview.

The criteria for inclusion in the series were as follows:

(1) All manifest deviations.
(2) All exophorias of 9 prism dioptres or more.
(3) All esophorias of 7 prism dioptres or more.
(4) All hyperphorias (including alternate sursumduction).

Results

PREVALENCE OF STRABISMUS

On the basis of the criteria used, the population contained 339 children with "abnormal" cover tests (7·1 per cent.). The prevalence was slightly higher for boys (7·3 per cent.) than for girls (6·9 per cent.), but the difference is not significant. The more detailed orthoptic examination was carried out on 319 of these children (94 per cent.), and on a control group of 310. The proportion of 7·1 per cent. includes all types of strabismus and large heterophorias, and is therefore relatively useless until we know the contribution of the various diagnoses to the total prevalence. This is given in Table I.

Table I *Relative prevalence of various types of squint*

Type of squint		*Percentage of all abnormal cover tests*	*Prevalence per thousand*
Exophoria 9Δ or more		9·4	6·7
Esophoria 7Δ or more		9·4	6·7
Divergent	Intermittently manifest	7·0	5·0
	Manifest	1·5	1·0
	Consecutive	2·4	1·7
Convergent	Fully accommodative	9·4	6·7
Convergence	excess	5·6	4·0
	Partially accommodative	21·5	15·3
	Non-accommodative	14·5	10·2
Other various diagnoses		13·3	9·4
Not examined in detail		5·9	4·2

The relative contributions of each type of case are, I think, what most of us practising orthoptics in this country would expect. Unfortunately, comparisons of these figures with those published by other authors is difficult, either because the other data are drawn from inadequately defined or non-comparable populations or because the diagnostic criteria are too crude, cases often being classed simply as "convergent" and "divergent or "constant" and "intermittent". However, one American study (Fletcher and Silverman,

1966) of 1,110 consecutive cases attending a clinic uses diagnostic criteria which make comparison of relative frequencies possible. The nature of this study was such that it is impossible to define clearly the population from which the cases were drawn, and overall prevalences are therefore unknown. Table II shows the relative frequencies where com-

Table II *Comparison of relative prevalences*

Type of squint		*Prevalence per thousand*	
		Cardiff	*Fletcher and Silverman* (1966)
Divergent	Intermittently manifest	9·4	18·7
	Constant	2·0	1·0
	Consecutive	3·1	0·7
Convergent	Accommodative	12·5	9·7
	Partially accommodative	36·1	28·2
	Non-accommodative	19·2	23·6
Other diagnoses		17·6	18·1

parison is possible. Exophoria and esophoria are omitted because diagnostic criteria are known to differ, and as the American study did not distinguish between partially accommodative and convergence excess types of squint, these two groups have been amalgamated.

The two series show a good overall correspondence with the exception of intermittent divergence, which is twice as common in the American group. This difference is highly significant ($P<0.01$). This marked difference in the relative frequency of intermittent divergence confirms a clinical impression quite widely held, and is a point worthy of further epidemiological study.

Overall prevalence figures may reasonably be compared with those of Frandsen (1960), who found a prevalence of 6 per cent. of manifest squint at the age of 6 years in Copenhagen in a comprehensive survey. The comparable figure for the Cardiff population is 5·33 per cent. for confirmed cases (this can be corrected to 5·66 per cent if an allowance is made for those cases suspected at the initial examination but not seen for full orthoptic examination). The two seem to agree surprisingly well.

Having defined the prevalence of squint in children aged 5 to 6 years, it is of interest to examine the manner in which this total prevalence developed. In the published literature there is a rather surprising divergence of opinion. The earlier British writers (Smith, 1899; Worth, 1906), held that the maximum incidence of convergent squint occurred at around the age of 24 months. Studies by Nordlöw (1953, 1964), Keiner (1951), and Scobee (1951), however, found that a majority of convergent squints arose before the age of 12 months, and these workers concluded that the previous findings were incorrect. In a comprehensive Danish survey, Frandsen (1960) again reversed the position, finding that only 15 per cent. of squints arose before the age of 12 months, and 50 per cent. in the second and third years of life.

The figures from Cardiff confirm the original British view—and I think our own clinical impressions. Of all squints, 22 per cent were found to have a stated onset before the age of 12 months and 43 per cent. in the second and third year. The median age at onset for all squints is 29 months. These figures, like overall prevalence, are rather crude, and normal clinical practice makes us well aware that the age incidences of the common types of squint are different. Referring to the three common varieties of convergent squint only, there is a notable difference between the fully accommodative, partially accommodative, and non-accommodative varieties, these having in the Cardiff study median ages at onset of 40, 30, and 21 months respectively.

These figures can be compared, because of the similarity of the classification, with those of Fletcher and Silverman (1966), whose quoted data give, on calculation, median ages at onset for partially accommodative convergence of 19 months and for non-accommodative of 13 months, both considerably earlier than in Cardiff. One difference between the two groups which may be relevant is that all histories in the American series were probably obtained at the time of presentation for treatment, whereas many of the Cardiff histories were taken at the age of 5 to 6 years. Although it would seem likely that the later histories would be less accurate, the only evidence available about the nature of possible bias related to the time interval seems to be the finding of Nordlöw (1953), who claimed that his study showed that the longer the interval between onset and history taking, the earlier the onset tended to be placed. Correction for such bias would exaggerate rather than decrease the difference between our figures and those of Fletcher and Silverman.

The later onset of accommodative squints in both series (very striking in fully accommodative cases, with only 15 per cent. said to have arisen before 2 years, as compared with 63 per cent. of non-accommodative squints) accords well with accepted views on the role of accommodation in the genesis of strabismus. The apparently contrary findings already quoted must be explainable either by true differences in relative prevalence, with a much higher proportion of non-accommodative squints, or by gross differences in definition and sampling procedures. As no detailed diagnostic categories for these workers are quoted, there is at present no means of telling whether the difference lies in technique, and is therefore of no interest, or represents a true difference in incidence which requires investigation.

Factors influencing prevalence

The list of factors which are believed or known to influence the prevalence of squint is a long one and time permits the discussion of only one particular aspect of this problem. It has long been believed that there is a hereditary element in the aetiology of many cases of squint, although varying opinions have been expressed as to the nature of the mechanism concerned. The view that the undoubted tendency of squints to run in families is of genetic origin is so widely held that it is surprising to find that, in discussion of the results of a survey of children in Newcastle (Miller, Court, Walton, and Knox, 1960), it was considered that the familial tendency should be explained in terms of environment rather than of heredity, as the observed pattern of inheritance "cannot be explained in terms of orthodox genetic theory". This latter statement is not correct, however. As Grützner, Yazawa, and Spivey (1970) have pointed out, the pattern of inheritance observed in strabismus, and in particular the high concordance in identical twins and much lower concordance in dissimilar twins, is compatible with polygenetic or multifactorial inheritance.

Our own study was not designed with a view specifically to elucidating the genetics of

strabismus, but it does, as expected, show that there is a tendency for squints to run in families. Comparing the children having manifest deviations with controls, there is a history of squint in a parent or sib, or both, in 12 per cent of the control series and in 25 per cent of the children with squints. Rather less reliably, 8·1 per cent. of controls gave a history of squint in a relative other than a parent or sib, as compared with 19 per cent of the squinting group.

The study of our own figures on family history is of little value in elucidating the mechanism of inheritance, but it is rewarding and interesting to examine in the light of our data the hypothesis advanced by Miller and others (1960) that the familial tendency is environmental rather than genetic.

It was shown by Thompson (1922) that in Glasgow at that time there was a highly significant variation in the prevalence of squint between four different social groups as defined by the school attended. This observation has been quoted many times in subsequent literature and is often cited as the authority for the statement that social class influences the prevalence of squint. The more recent study in Newcastle already quoted and the National Child Development Survey (Davies, Butler, and Goldstein, 1972) "confirm a trend" towards such variation, but give only Thompson's data in support, quoting no figures of their own.

The Cardiff study fails to show any relation whatsoever between social class and the prevalence of strabismus. Table III shows the proportion of the squinting and control

Table III *Prevalence of manifest squint, by social class (per cent.)*

Social class	*I*	*II*	*III*	*IV*	*V*
Cases of manifest squint	8	13	54	12	13
Controls	9	15	50	10	15

series which come within each of the five groups of the Registrar General's Classification. What is striking about it is the similarity of the distributions, suggesting that in respect of social class the squinting series is a random sample. Further analysis of the data by differing types of squint fails to reveal any significant correlation concealed by the aggregation of all squints into a single group.

This result seems to conflict directly with that of Thompson. The most notable differences, however, are not those between the correlations themselves, but the interval of almost 50 years between the two observations, and the marked difference in prevalence: 5·7 per cent. for manifest squint in Cardiff today and 2·9 per cent. in Glasgow in 1922. In this 50-year period the rise in the standard of living of social classes IV and V has been very much greater than that in the other classes. Glasgow in 1922 had the unenviable reputation of having the worst housing in the United Kingdom, and the unskilled manual worker in that time was vastly worse off in almost all material respects than his modern counterpart in Cardiff. Thompson's four classes do not correspond with the five classes currently used, but a study of his quoted figures shows that the differences between his classes I, II, and III just fail to reach significance at the 0·05 level, while the difference between these three and Class IV is very highly significant. It could well be that the modern equivalent of his Class IV represents only a negligible proportion of Class V under the current system and that

the increased prevalence he observed in the lowest class is seen only at standards of living fortunately uncommon in modern Britain.

If, as appears to be the case, squints were more frequent in the lowest social group in Glasgow in 1922 than in Cardiff today and if, as seems reasonable, the prevalence in other classes is at least no higher today than in 1922, Thompson's estimate of prevalence should have been higher, not significantly lower, than that now seen in Cardiff. This rather suggests that the Glasgow Survey may have failed to detect the less cosmetically obvious squints, a point which is further discussed below.

In both these studies, social class has been used as a rough guide to the child's environment as determined by a number of factors. In the Cardiff Study some of these factors were defined separately and an attempt can be made to assess their individual importance.

HOUSING

The standard of housing was indicated in the records by an index derived from the ratio of rooms to occupants. Using this index, there was no significant difference between any of the types of squint and the control group (Table IV).

Table IV *Standard of housing, by ratio of rooms to occupants (per cent.)*

$\frac{Rooms}{Occupants} \times 5$	1	2	3	4	5	6	7	8	9 or over
Squint	0	0	1	9	14	18	20	10	27
Control	0	0	1	9	11	19	20	8	31

FAMILY RELATIONSHIPS

Neither the number of sibs (Table V) nor the position of the squinting child in the sibship (Table VI) appears to be related to the appearance of squint, nor did any squinting group show a higher proportion of children falling in the composite group of illegitimate children

Table V *Relation of squint to number of sibs (per cent.)*

No. of sibs	0	1	2	3	4	5 *or over*	*Not known*
Squint	6	31	33	15	8	7	0
Control	7	29	26	18	11	8	1

Table VI *Relation of squint to position of child in sibship (per cent.)*

Position in sibship	*1st*	*2nd*	*3rd*	*4th*	*5th*	*6th*	*Institutional care*
Squint	32	32	16	9	5	3	2
Control	34	25	19	11	5	3	3

and those who were adopted or whose parents were divorced, the proportions being 6·85 per cent for strabismus cases and 6·90 per cent. for control cases.

STANDARD OF CARE AND NUTRITION

This is hard to assess with accuracy, but an attempt was made to grade children into four categories according to the general state of their clothes, tidiness, and cleanliness. This assessment revealed no significant difference between any type of squint and the control group (Table VII). This is in contrast to the findings in Newcastle (Miller and others, 1960), where there was a significant difference, with a prevalence of 8·2 per cent. of squints in children whose care was deficient compared with 3·9 per cent. in the remainder of the population. It is perhaps of interest to note, however, that in the Cardiff series the results of treatment of all types of convergent squint are significantly worse in those children who appear to show a poor standard of care (Table VIII).

Table VII *State of care of child in squint and control groups (per cent.)*

State of care	*Good*	*Indifferent*	*Poor*	*Very poor*
Squints	70	22	6	2
Controls	67	23	10	1

Table VIII *Result of treatment (percent.) according to state of care in 173 cases of convergent squint*

State of care	*Good*	*Indifferent*	*Poor*	*Very poor*
Result satisfactory (67)	82	13	3	1
Unsatisfactory (106)	58	29	8	4

$0{\cdot}05 > P > 0{\cdot}02$

The frequency of childhood infections, which are often suspected of being a precipitating factor in the onset of strabismus, might be expected also to correlate with environmental conditions and social class, and on both grounds it might be expected that children with squints would show a higher incidence of such infections. Such an association has been reported by Frandsen (1960), but it was not found in the Cardiff series (Table IX) or in the Newcastle Study.

Table IX *Frequency of childhood illness in squint and control groups (per cent.)*

No. of "childhood illnesses"	0	1	2	3	4	5 *or over*	*Not known*
Manifest squints	13	37	29	15	4	1	1
Controls	17	34	31	14	3	0	0

It seems quite possible that a number of previous studies, where the initial screening at least has been carried out by staff with no special ophthalmological or orthoptic experience, may have detected only the cosmetically obvious squints and that an overall correlation of the presence of squint with the standard of care and the apparent association between high prevalence and social class has its origins partly or wholly in the fact that such surveys fail to detect a higher proportion of manifest squints than those carried out by personnel engaged in the daily practice of orthoptics.

To summarize this brief review of the apparent effect of social conditions, it would appear that in Cardiff today social class has little if any effect on the incidence of squint. There are, however, indications in the literature which suggest that strabismus is more frequently seen, especially in its more cosmetically obvious forms, in those who live at low environmental standards much less common today than in former years. I would add that those classed in the Cardiff study as showing poor care are by no means confined to the lower end of the social scale. The principal importance of poor care and environment under current conditions appears to lie in the fact that such children are more difficult to treat effectively rather than that they more commonly develop strabismus.

Finally, our findings offer no support to the hypothesis that the familial tendency in strabismus is environmental, having its origin in social conditions. It seems much more probable that it can be explained as being the result of the unfortunate coincidence in one individual of several independently inherited genetic factors.

All epidemiological work of this sort is the result of a team effort and many people in the Medical Research Council's Epidemiological Research Unit and the Health Department of the Cardiff City Council have, by a great deal of hard work, made it possible to obtain and present these results. It would be impossible to name them all, but I would particularly like to mention Mrs. E. Childs, the Research Orthoptist who carried out all the very detailed examination and interview work and assembled the results, and Prof. A. L. Cochrane, Director of the M.R.C. Epidemiological Research Unit, for his advice and encouragement.