17

Discussion: Epidemiology of Blindness

I K Jalili

17.1 The Survey

Between 1985 and 1987, 709 persons were registered in this study and over 2500 related persons were screened. This was undertaken by the author who was based at the SJEH, Jerusalem, during this period. This hospital was the main ophthalmic centre for the whole of the Palestinian population in both the West Bank (WB) and the Gaza Strip (GS).

The patients comprised a wide array of ages and backgrounds that fulfilled the criteria of childhood onset visual impairment in the WB and the GS and included examining the pupils of the 5 blind schools and training centre in the two Palestinian regions). In the GS, the survey was extended to include home visits to families who were suspected of having children with visual impairment. The restricted geographical area and the demographic pattern of the GS, where the bulk of the population were concentrated in confined locations within towns, camps and villages with an easy access by car, made such comprehensive screening possible. This was combined with an excellent input by the dedicated social worker at the UNRWA School for the Blind in Gaza, without whose knowledge and assistance, such an elaborate study would not have been possible.

The study has also highlighted the patterns of many rare conditions that existed in large pedigrees with several affected siblings and also enabled the evaluation of the natural history of these conditions and their intra and interfamilial variability. The size of the cohort and the comprehensiveness of the protocol in this geographically and socially confined population made it possible to encompass virtually all the visually impaired children, especially those in GS, thus eliminating significant bias normally encountered in such population surveys. [(1), (2), (3), (4)]

17.2 Possible Bias

The possible source of bias in this study could have resulted, in both the school age and adult population, from: -

Bias in the School Age Cohorts

1. Some under representation of those with albinism, particularly in the WB; many of whom were educated in sighted schools and the author of this thesis has come across a few such cases.

2. Several cases (both children and adults) with congenital nystagmus in the GS declined to be examined or enrolled in the study.
3. The presence of female only residential primary schools in the WB and in close proximity to Jerusalem and the male only, essentially non-residential school in Bethlehem, could have influenced the gender per age distribution in the WB and possibly limited the number of younger males in the study.
4. A small under representation of the pre-school <6 age cohort in the following: -
a. Some young children, particularly surgical cases, whose families are covered by the Israeli health insurance system, especially from Eastern Jerusalem and possibly, but to a lesser extent, from the GS might have filtered to the Israeli ophthalmic services.
b. A number of cases who did not have existing relatives in the blind schools in the WB could have been kept at home until they reach school age. Such cases, if they exist, must have been seen at SJEH at some stage and included in the study through the outpatient's casenotes cohorts.
c. A very small number of blind children whose families live in the WB who are sent as borders to blind schools abroad such as Jordan and, in the more affluent families, to blind schools in the West particularly Canada and the US where there are large Palestinian communities.
5. Only a few children with multiple handicaps were encountered in the three residential homes, two in the WB and one in the GS. It is possible that such children were kept at home and looked after by their families, and it is highly probable that the more severe and weaker patients die as a result of their afflictions and lack of care.

Bias in the Adult Cohorts
The possible source of bias in this cohort could have resulted from:
1. An unknown proportion of adults especially males, both in the WB and GS who did not present to the survey, because of work commitment, may have been underrepresented in the study. Fieldwork carried out at the weekends, however, must have, however, reduced the size of this problem especially in the GS. In the WB we were informed at the start of the study that only very few blind people were present in one or two workshops in Jerusalem and all had adult onset blindness.
2. An additional 83 adult patients were not available for examination. This number has been added to the figures obtained from the study when the estimates of the prevalence of blindness were calculated.
3. A bias in the GS has been introduced as a result of a more comprehensive coverage helped by the smaller geography and a closer support from social services.

17.3 Patients and Schools

The blind in the OPT were educated in special blind institutions. The educational system, especially in the WB, was not systematically structured and lacked methodical planning because of the absence of a national authority combined with prevailing poverty and limitations of resources in the community.
The service was very influenced by religious charities, both local and international. This was noticeable with regard to girls' education in particular. Apart from a small school and training workshop in Nablus, all institutions were based in the central region of the WB in close proximity to Jerusalem. There was only one governmental school that catered for boys for both primary and secondary education; again, this was situated centrally in Bethlehem.

In the northern city of Nablus a small school and training workshop provided the service and ensured proximity of the blind children to their families in the north of the country.

The remaining centres were essentially residential homes, also run by charities, catering for all disabilities and providing some support for the very young and adults pursuing higher education in the WB.

In Gaza City, there was one major non-boarding primary school and training centre catering for the visually impaired population of the GS which was supported by UNRWA. Secondary school education was provided at

sighted schools with a tendency for the blind to enrol in educational institutions run by religious establishments such as Al-Azhar Institute in Gaza.

17.4 Sex Ratio

Two distinct patterns are demonstrated in the proportions of males and females in the study; one influenced by patients' demography, i.e. the WB and the GS; and the other by the conditions' aetiology, that is whether hereditary or non-hereditary.

Sex Ratio by Region

Male preponderance in the total series (1.44:1) is influenced by an excess of males in the GS cohorts with 176 males and 114 females giving an M:F ratio of 1.54:1 (Chapter 12, Table 12.1). This predominance is echoed in all age cohorts, although it peaks in the < 5 and 16-20 years cohorts and become less noticeable in the school age group. It is possible that the difference is a selection bias when it comes to competition for limited spaces in the < 5 cohort and a less inclination of the blind teenage girls to come forward, especially in the absence of any hope for a cure.

In the WB, however, there was an almost equal proportion of both genders with 172 males (49.5%) and 175 females (50.5%) giving a sex ratio of 0.98:1, almost identical to that of the general population. Numbers and ages are as at the end of the study.

In the school age group, the gap in the sex ratio diminishes reaching near normal population ratio and ranging between 1:1 to 1.2:1 depending on the criteria used in extracting the data; that is whether the age range is taken using the age at examination, pupils only, or all the children in that age range. In the school age cohort, there is an equal representation of both genders in the study, especially in hereditary cases, identical to that of the M:F ratio in the general population. The small numbers of these two categories, together with the almost absent X-linked conditions in the series (0.57%), did not skew the sex ratio in the total series towards male preponderance as is the case in several other studies, especially those based on school surveys and blind registers. [(5), (6), (7), (8), (9), (10), (11)] Similar to these studies, optic nerve and prenatal conditions caused the male preponderance in the non-hereditary conditions.

The other pattern noted is the presence of a male preponderance in the < 5 and 16+ age cohorts in the GS which is reversed in the WB (more pronounced in the 16-20 years group). (Chapter 12)

In the < 5, the predominance of males in the out-patient cohorts in both regions suggests male favouring in seeking medical advice; however there in no solid evidence to support this conclusion. The reverse finding among the non-outpatient cases in the WB is caused by the availability of female-only primary and residential schools in the region. In the 16+, the predominance of males is hard to explain. To the best of the author's knowledge, and apart from a few cases, both sexes were equally represented in the series. The high mortality rate of females in the sibships studied (0.43:1) is not matched by a correspondingly high mortality in the affected patients (0.92:1, n=25) nor in the general population where there is a higher rate of male deaths in both regions, although, figures on the GS alone are not included in the references used. [(12)]

This study also reveals the absence of a culture favouring males in education as witnessed in some developing countries. If anything, there is a slightly higher presence of women in the WB cohort as a result of the females' only training workshop in the region. This similarity in the gender ratio was also demonstrated by Elder and De Cock but without a breakdown of their results between the two Palestinian regions. [(13)] The few cases of children in the school age who were found on the waiting list are from both genders and the delay in registration was caused by the remote location of their villages.

Table 17.1 Comparison of M:F ratios with worldwide data

Country	Sample	M:F [a]
This Study (whole series) [b]	669	1.22:1
WB	347	0.96:1
GS	289	1.56:1
This Study (< 16) [c]	405	1.29:1
WB < 16	214	1.12:1
GS < 16	164	1.45:1
WB and GS	205	0.88:1
Lebanon	231	1.7:1
Jordan	260	1.8:1
Iraq	150	2.5:1
Egypt	113	2.8:1
Saudi Arabia	372	1.45:1
Uzbekistan	671	1.44:1
Nordic Population	304	1.4:1
Iceland	43	2.6:1
Netherlands	1334	1.6:1
Bavaria, Germany <20	1141	1.2:1
Czech Republic	229	1.76:1
UK (National active surveillance)	439	1.17:1
Ireland	172	1.18:1
Chile	267	1:0.97
Peru	202	1:0.96
Thailand	127	2.63:1
India	305	1.7:1
India (in 1968)	180	12.8:1
India (population based survey)	5	1:0.66
Tanzania (hospital based study)	130	1:0.92

[a] M:F population ratio is 1.1:1.
[b, c] Includes additional 33 and 23 cases respectively to the WB and GS figures

Sex Ratio by Condition

In the non-hereditary and undetermined groups, there is a male preponderance of 1.8:1. The former is caused by optic nerve conditions (n=8, 100% males) and acquired (non-traumatic) cases (6 males, 2 females).

This study has demonstrated that all blind children in the region attend blind schools without any gender bias. This is unlike many other developing countries where only a small minority attends blind schools. In India for example, only 10% of blind children attend schools for the blind, and these are predominantly male. [(14)] In the WB and the GS there is also an obvious desire by the families to send their children to the blind school for better education and a better life even in the most deprived, uneducated classes and families in remote areas. This has been greatly enhanced by the free boarding facilities, sponsored by charities in the WB, and by UNRWA in the GS, available in several schools and institutions in the WB, especially at the level of primary education. All trainees in the workshops are female and training provides them with the means of being breadwinners after leaving the centre. It is interesting to note that in Jordan, where 60% of its population are Palestinians, [(15)] a male preponderance of 1.8:1 was reported and was attributed to the prevalence of X-linked conditions, increased exposure to trauma, and social traditions favouring males in some communities. [(16)], [(17)]

17.5 Pedigrees and Sibships

The identical ratio of pedigrees in relation to the general population in both the WB and the GS indicates a similar incidence of pathologies in both regions and hence the mutation rate, particularly as genetic conditions formed the majority of cases. There are however larger numbers of sibships per pedigree, and consequently patients, in the GS giving rise to a higher prevalence of childhood onset disorders. This has been caused mainly by the predominance of conditions such as CRD and microphthalmos in that region.

17.6 Visual Acuities and Prevalence

The spectrum of visual acuities in the series ranges from NVI to NLP. (Figure 17.1)
Category '1' (NVI), as it represents acuity better than 6/18, is not usually reported in blind school surveys. In this study it averages 6% (WB 6%, GS 10%) with almost one fifth of the total category having significantly compromised vision in the other eye. Two thirds of cases in this category are of school age and attending blind schools. The presence of such cases with reasonably good bilateral visual acuity in the blind schools is explained by; (a) the absence of any assessment of these children prior to their admission to the schools; and (b) the flexible criteria in admitting pupils to these schools in both regions. Over three quarters of the pupils in this category could have been taught in normal sighted schools, although about a third have progressive conditions such as RCD and CG and needed to be prepared for the future in blind schools. The percentage of this category in the total series is similar to parts of the world such as Uzbekistan 6.1%, [6] and Chile 6.1%, [18] but is much lower than the Jordanian study where nearly 24% of the series fell into this visual category. [5] This would indicate that the same trend, but to a greater degree, is present in Jordan.

There is also a preponderance of patients in category '2' (VI) in the GS at 42%, (WB 34%) as a result of the higher prevalence of cases with achromatopsia, albinism and CRD. This category is the second largest visual acuity category and a large proportion of those affected are in the < 16 cohort. The proportion of this category in this study is second only to the Nordic population. This can be explained on the same basis as in category '1' and has only been reported in some of the studies. [5], [6], [18], [19], [20], [21], [22], [23] These conditions are overlooked when data are extracted from blind registers. [22]

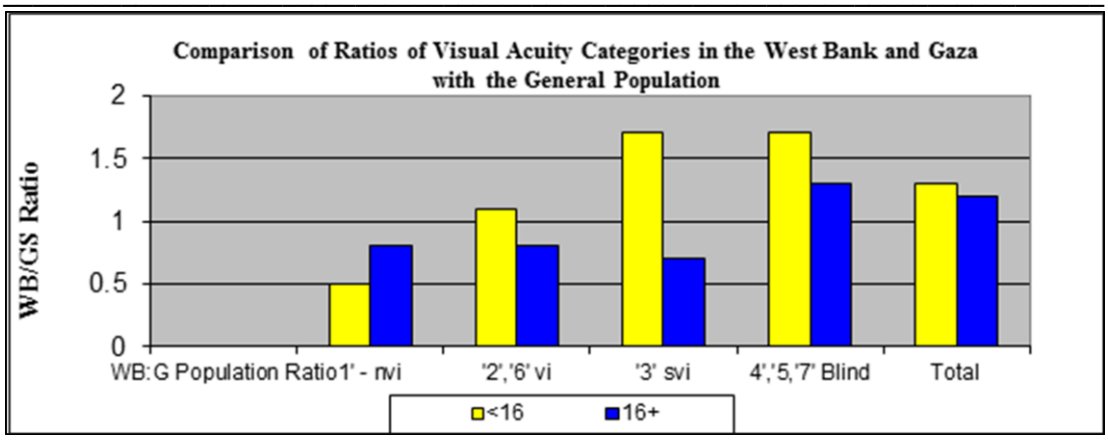
The SVI category '3' was small at 9.4% (WB 11.2; 19.5%), and is similar to percentages in other studies. [6], [18] [19], [21], [23], [24] Analysing the range of visual acuities in this category reveals a severely compromised group of people with over a third (37%) with LP and 38% with CF and HM acuities forming 75% of the range of vision in this group (Table 17.2).

Blindness (categories '4' and '5') formed 46 of the total series. The disparity between the two regions is found in both adults and children (WB 51% and 42%, GS 41% and 32% respectively) and is the result of higher numbers of cases with congenital CRD in the WB. The average figure for the SVI category in both regions of 39.8%, which is similar to the figure in the Saudi study, [20] is lower than the figures given from the same region in the study carried out six years later in which the SVI category forms 63% of the total. This is the result of the lower proportion of categories '1' and '2' in that study, (Table 17.2) which suggests the higher figure of SVI in the later study is the result of lower proportions of categories '1 and '2' in their survey, and that the recommendations put forward by the author of this thesis in 1987 had been implemented and more children with reasonable visual acuities were being taught in sighted school. In fact stricter admission criteria began towards the end of this survey based on the reports submitted by the author of this study to Social Services and Educational Authorities.

The combined SVI/BL forms 62% of the total series and ranged between 60% in the WB, to 47% in the GS in the < 16. This exceeds the figures from Jordan which were 28% in those born before 1970, and 38% in those born after 1970 with an average of 33%. Therefore, our figures take a middle position in comparison to the data from the rest of the world. [5], [6], [18], [19], [20], [21], [22], [23], [24], [25], [26]

Figure 17.1 highlights the pattern of visual acuities in the rod-cone and cone series where 44 patients with acquired conditions are in SVI/BL categories. This is a substantial number considering that there are only 55 acquired cases thus forming 80% of the total acquired conditions. The same proportion applies to the microphthalmia/anophthalmia group with a total number of cases in the series of 30. The majority of those have NLP. These figures exclude the microphthalmia associated with cataract, and are classified accordingly.

Figure 17.1 Categories of visual acuities by region



Congenital glaucoma is another condition characterised by severe visual disability and blindness and an extremely poor prognosis. Out of the total number of 68 cases with CG, 35 (68%) fell into this combined visual category and two thirds have NLP. The two major types of RD, namely CRD and RCD, exhibit their severity and prognosis by their acuities whereby CRD, with the better prognosis, are represented with only 42% of the total series in the SVI/BL groups with only one patient with NLP and this in a man of 50 years of age. In contrast, patients with RCD formed 79% of the total series of (n=90/114) in this combined category with 10 patients with NLP. The majority of optic nerve disorders (n=17, 89% of the total cases) fall into this category with 7 and 10 cases in categories 4 and 5 respectively. The cases with myopia syndromes share 38% (n=10) of the combined category but 9 out of 10 have NLP. Congenital cataract has only 18.6% share of this group with 25 cases; the majority of whom fall within the SVI category. [6], [27], [26], [28], [29] [30], [31], [32], [33], [34], [35], [36], [37], [38]

Despite the disparities in the percentages of blindness between the WB and GS, the prevalence of SVI/BL in the two regions in children is equitable at 0.28/1000 (Table 17.2) The prevalence of blindness in all age groups is even lower at 0.19/1000 populations. The prevalence in all ages is different in the two regions (WB 0.24/1000, GS 0.30/1000) indicating a higher prevalence in the 16+ in the GS than in the WB. These figures are surprisingly low taking into account the level of consanguineous marriage in the region. They fall within the range of those reported in Europe such as in the Nordic countries, Ireland and the UK, [8], [9], [28], [29] rather than those found in Asia and Africa where the highest figure reported is in Malawi (1.1/1000). [39], [30], [31], [32] This low prevalence in the WB and GS, which is also applicable to other countries in the region such as Lebanon; 0.1/1000 can be explained by; [7] (a) the predominance of conditions that cause blindness at a later age, especially retinal dystrophies; (b) the absence of perinatal causes such as those related to prematurity which are predominant in industrialised countries; and (c) the absence of nutritional and infective causes of blindness widely seen in Africa and some parts of Asia.

Table 17.2 Distribution of visual acuity categories (%)

Visual Acuities →	Ages	NVI 6/18 + 6/60	VI <6/18-6/60	SVI <6/60-3/60	Blind <3/60-PL	NLP	SVI/BL <6/60-NPL	UD	Reference
Middle East and Uzbekistan									
West Bank and Gaza Strip									
WB and GS (n=296)	5-15	7.6	29.9	11.5	39.78	10.2	56.2	0.7	This study
WB and GS -(n=669) [a]	0-86	5.7	31.2	9.6	46.3	16.1	61.7	1.3	
WB (n=347)	0-77	3.7	28	10.1	51	14.9	67	6	
GS (n=289)	0-86	8	35	10	41	15.5	56	2	
WB and GS < 16 (n=405) [a]	0-15	6	38	9	38	12	55	1	
WB < 16 (n=214)	0-15	4	34	11	42	14	60	1	
GS < 16 (n=164)	0-15	10	42	9	32	10	47	2	
WB and GS 16+ (n=264) [a]	16-86	4.9	21.2	9.8	59.8	20.8	72.7	1.1	
WB 16+ (n=133)	16-77	3.8	18	8.3	66.2	20.3	76.7	1.5	
GS 16+ (n=214)	16-86	4.8	25.6	12	52.8	21.6	68.8	0.8	
WB and GS (n=205)	5-15	2.9	12.7	21	63	-	84	-	Elder & De Cock (1993)
Jordan (n-260) Before 1970	0.8-47	43	27	13	15	6	28	-	
After 1970		44	19	21	17	7	38	-	Al-Salem & Rawashdeh (1992)
Total		43	24	16	17	7	33	-	
Lebanon (n=203)	0 - 20	-	-	60*	84	40	100	-	Baghdassarian & Tabbara (1975)
Saudi Arabia									
Riyadh (n=187) *	Mixed	-	-	-	-	31	100	-	Tabbara & Badr (1985)
East Province (n=219)	6 - 60	-	3.2	4.1	48.4	43.8	52.5	-	Badr & Qureshi (1983)
Eye referral study (n=372)	2 - 18		70		30	-	-	-	Tabbara *et al* (2004)
Iraq (n=150)	< 12	-	-	-	-	32	95	-	Alkanani (1990)
Egypt (n=113)	6 - 19	-	-	-	-	10.6	89.4	-	El-Gilany *et al* (2002)

Uzbekistan (n=671)	0 -15	5.1	18.9	16.1	59.3	-	75.4	0.6	Rogers *et al* (1999)
INDIA AND SSA									
South India (n=329)	3 - 28	2.5	5.8	6.7	86	-	92.7	-	Gilbert *et al* (1993)
West Africa (n=315)	3 - 28	1.9	7.9	6.6	83.6	-	90.2	-	Gilbert *et al* (1993)
Nigeria (n=142)	Mixed	0	1.4	2.8	54.3	41.5	98.8		Ezegui *et al* 2003
LAC (Latin America and the Caribbean)									
Chile (n=318)	0 - 15	6.6	9.4	9.2	74.8	-	84	5	Gilbert *et al* (1994)
Chile (n=261)	3 - 28	6.1	10.7	9.2	74.4	-	83.6	-	Gilbert *et al* (1993)
Peru (n=202)	0 - 18	-	-	14	86	-	100	-	Rojas *et al* 1990)
Jamaica (n=108)	5 - 15	-	-	-	-	-	100	-	Moriarty (1988)
Europe, North America and Australasia									
Denmark (n=150)	0 - 18	-	36	-	-	-	39.3	24.6	Rosenberg (1987)
Nordic Population (n=304)	0 - 17							-	
Brain Group (n=135)		15	12	19	17	-	36	-	Rosenberg *et al* (1996)
Ocular Group (n=168)									
Nordic Study (n=2527) [b]	0 - 17	-	39.8	-	22	-	-	24.7	Hansen *et al* (1992)
Sweden	0 - 19	45					25		Blohme & Tornqvist 1997
Iceland (n=43)	0 - 15	46.5	-	46.5	-	7	43.5	-	Halldórsson & Björnsson (1980)
UK (n=728)	0 - 15	N/a	n/a	51.1	40.8	-	91.9	8.1	Evans *et al* (1991)
Scotland (n=99)	5 - 18	-	-	-	-	-	100	-	Phillips *et al* (1987)
Northern Ireland (n=486)	0 - 20	-	-	-	-	-	-	-	Bryars & Archer (1977)
Irish Republic (=172)	0 - 16	-	-	-	-	-	-	-	Goggin & O'Keefe (1991)
Australia (n=311)	Mixed	-	-	-	-	-	-	-	Yeates (1983)
Australia, South (n=50)	5 - 18	-	-	-	-	-	-	-	Fraser (1968)
Czech Republic (n=229)	6 - 15	-	-	20.5	69.5	-	90	-	Kocur et al (2001)

[a] Also includes additional patients of uncertain OPT region or from Israel. UD: Undetermined. [b] Both categories 3 and 4 combined

17.7 Site of pathology

Retina and cornea as sites of primary pathology form the ends of the scale, when the prevalence of one is high; the other tends to be low. Retina therefore is an important site in countries where they have achieved good standards of health care such as industrialised countries and many of the ME countries, reaching as high as 60.8% in the UK in the isolated SVI/BL cases. [6], [7], [9], [20], [40], [41], [42], [43] Percentages of retinal conditions rise sharply when genetic conditions are analysed separately, a feature clearly seen in countries with a high prevalence of anterior segment disorders secondary to infections and malnutrition such as Malawi where its proportions jumps from 0.7% of cases combining all aetiologies, to 50% of the genetic cases. [44], [45] In the WB and GS, retina takes 44.8% to 51% of cases, depending on the region and age cohort, being the highest in the GS in the < 16. The high prevalence in both Palestinian regions was also confirmed in a later study at 52.1% in 1993. [27] The Palestinian figures are amongst the highest in the world and are significantly higher than the rest of the Arab countries which range between 29.5% and 30% in neighbouring Jordan [5] and Uzbekistan [6] to 32% in Saudi Arabia [20] and 39% in Lebanon and Cyprus. [7], [40] The predominance of retinal conditions is caused by the high prevalence of retinal dystrophies and, in the middle-income countries, by ROP. [9] (Table 17.4)

The second anatomical site in importance, especially in children, is the lens which forms one quarter of cases in the total series as depicted in Table 17.4. The lower percentage of lens conditions in the 16+ in the WB is most likely the result of under representation in comparison to the better coverage of the corresponding age cohort in the GS. These figures come third to Saudi Arabia 34% [20] and Uzbekistan 30.2%. [6] They are higher than Jordan 20% and Lebanon 18%. [7] The preponderance of lens conditions in the MEC is the highest in the world and is the result of the high rate of consanguinity in this region, which are several folds higher that figures from EME of 4.8% to 8.7%, [41], [42], [43], [46] and, 7 to 14% of genetic cases in Denmark and Iceland; and most regions in Africa. [18], [34], [45], [47], [48] with the exception of Nigeria. [24], [49]

WHO classification of anatomical sites incurporates glaucoma in the whole globe category, but recent publications address this pathology separately. According to the original classification, the whole globe ranks third in the total series. In the WB, its prevalence in the 16+ is double that of the < 16 cohort (26.3%, 13% respectively). The proportion of this category in the WB and GS are similar to the rest of the MEC and India which range between 17–23.1%. [5], [6], [7], [18], [20], [27], [40] The prime conditions in this category are buphthalmos (10.6%) and microphthalmos (5.5%); which compare well to the findings 6 years later (buphthalmos 9%). [27] (Chapter 19)

Optic atrophy is the sole pathology affecting the optic nerve which is a small category comprising 23 cases, ranking fourth among the anatomical sites. The significant disparities in the numbers affected between the two regions (WB 20, GS 3) and between age cohorts (children 18, adults 5) are hard to explain. The male preponderance in optic atrophy (1.5:1) is in line with that reported in other studies. However, the aetiologies of the atrophy is different, being predominantly due to childhood causes in this study unlike other studies such as the Nordic study where perinatal factors are the main causes. [8], [10] The proportion of optic nerve involvement in the 5-15 age cohorts is less than half of the 12% figure reported later from the same region. [27] (Table 17.4) In the rest of the world, optic nerve pathology varies considerably from as low as nil in some Arab countries, 1% in the GS, and 1.3% Thailand, to much higher figures in the EMC countries; the highest being the European and American rates of 23%, Denmark of 27.3% and the Czech Republic of 15.3%. Optic nerve conditions have become the leading cause of childhood blindness in developed countries.

Corneal conditions are the least important category and come fifth in the ranks of anatomical sites with marked differences in their proportions in children and adults. They are over three-fold higher in adults in the WB (7.5%

versus 1.9%) with no single case seen in children in the GS. This clearly demonstrates a change in the causation of disease over the years where bilateral visual disabilities from infections have come down significantly, and there has been a further improvement in the already low prevalence in adults.

Table 17.3 Worldwide prevalence of childhood blindness

MIDDLE EAST AND MUSLIM COMMUNITIES	
Palestinian Territories	
This survey <16	
WB	0.28
GS	0.28
This survey All ages	
WB	0.24
GS	0.3
5-18 (1993	
WB	0.17
Gaza Strip)	0.16
OPT (1993)	0.17
Other Middle East	BL 0.1
Lebanon (1997)	
BL	0.1
VI	1.1
Oman (2002) 0.08% 0-14 age group	0.08 [50]
Uzbekistan (1999)	0.5
EUROPE	
UK (1988)	0.36
Ireland (1991)	0.2
Ireland (1980)	0.36
Iceland (1992)	0.15-0.41
Scandinavia (1991)	0.15-0.41
Hungary (1991)	0.4-0.45
ASIA	
India (1995)	0.7
Mongolia (2002)	0.16
Nepal	0.64
AFRICA	
Malawi	1.1
Gambia (1986)	0.7
Benin (1995)	0.6
Kenya (1990)	
BL	1
VI	2
South Africa (1997)	
Blacks	0.7
Coloured	0.5
Indian	0.3
White	0.2
Average	0.35

These figures compare to those reported in Europe such as the UK [42] and the Czech Republic [9] in contrast to the high figures from India and Africa where the highest recorded are in East Africa (72%) and Malawi (75.5%) [18], [36], [45], [47], [48], [51] These are the result of the rarity of bilateral corneal infection and the absence of nutritional deficiencies, in particular VAD, in the population under study. The size of the problem in Jordan and Saudi Arabia prior to the improvement in health care facilities, appears to have been more severe than in the OPT when compared to the adult population in this series. [5], [20]

The rest of the anatomical categories are less important. However, it is worth mentioning that idiopathic nystagmus, an AR condition, is predominantly a GS condition which has resulted from the presence of an extended pedigree with this condition. Uveal conditions on the other hand are purely encountered in the WB with no cases found in the GS.

Table 17.4 Comparison of percentages of anatomical site worldwide

	Cohort	Globe	Glaucoma	Cornea	Lens	Uvea	Retina	Albinism	O.N	ONL	GAN	UD [a]	Total
Region/Country													
This Study – West Bank													
< 16	214	13.1	8.4*	1.9	26.6	1.9	45.8	2.8*	7.5	0.9	0.9	1.4	100
16+	133	26.3	11.3*	7.5	10.4	0.8	51.2	1.5*	3	0	0.8	0	100
Total	347	18.2	9.5*	4	20.5	1.4	47.8	2.3*	5.8	0.6	0.9	0.9	100
This Study – Gaza Strip													
< 16	164	20.7	11.6*	0	22	0	51.8	7.9*	1.2	0	3.7	0.6	100
16+	125	24.8	9.6*	5.6	20.8	0	44.8	4.8*	0.8	0	3.2	0	100
16+	289	22.5	10.7*	2.4	21.5	0	48.8	6.6*	1	0	3.5	0.3	100
Whole Series													
< 16	405	17	10.6*	1	25.9	1	46.9	4.7*	4.4	0.5	2.2	1	100
16+	264	25.4	10.6*	6.8	15.2	0.8	47.3	3*	1.9	0	2.7	0	100
Total	669	20.3	10.6*	3.3	21.7	0.9	47.1	4*	3.4	0.3	2.4	0.6	100
Other Middle Eastern Countries													
WB and GS 1993	205	23.1	9*	2.9	6.9	0.6	52.1	*	12.1	0	2.3	0	100
Jordan 1992	191	4	15	5	20	1	29.5	6	1.5	0	8	10	100
Lebanon 1975	157	11	6	2	18	1	39	9	0	0	0	14	100
Saudi Arabia 1985	106	0	17	0	34	0	32	0	0	0	0	17	100
Sudan 2005	40	2.5	2.5	40	12.5	-	7.5	-	2.5	-	-	32.5	100
Cyprus (g) 1972	89	11	6	2	18	1	39	9	0	0	0	14	100
Uzbekistan 1999	671	15.4	4.7	5.7	29.1	5.5	30.2	0	6.7	2.6	0	0	100
ASIA/INDIA													

Table 17.4 Comparison of percentages of anatomical site worldwide

	Cohort	Globe	Glaucoma	Cornea	Lens	Uvea	Retina	Albinism	O.N	ONL	GAN	UD [a]	Total
South Asia 1992	-	-	5	33	7	1	20	-	5	29	-	-	100
South India 1998	172	25	4.2	11.1	15.3	0	22.2	-	16.7	2.8	2.8	-	100
South India 1993	305	20	3	38.4	7.4	2	22.6	-	5.6	-	1	-	100
India 2001	2283	25	3	27	11	5	22	-	6	1	-	-	100
India (9 states) 1995	1318	25.3	2.6	26.4	12.5	5.8	20.7	0	5.9	-	-	0.9	100
India (g) 1995	303	3.9	0	1.3	4.3	0.7	79.7	4.3	-	-	-	5.6	100
North India 2003	703	27.4	-	21.7	10.9	-	15.1	-	-	-	-	-	75
Sri Lanka (g) 1995	79	6.3	5.1	1.3	16.5	1.3	56.5	1.3	-	-	-	11.6	100
Thailand/Philippines (g) 1993	244	7.3	7.3	0	43.9	0	41.5	-	-	-	-	-	100
Khoa-Kaen (Thailand)	65	27.7	6.2	12.3	16.9	1.5	29.2	-	6.2	-	-	-	100
Manila (Philippines)	113	27.4	8.8	13.4	16.8	0.9	23	-	8.8	-	0.9	-	100
Baguio (Philippines)	31	22.6	0	54.8	9.7	3.2	3.2	-	6.5	-	-	-	100
Davao (Philippines)	35	28.6	2.9	42.9	11.4	5.7	8.5	-	0	-	-	-	100
Malaysia 2001	332	17.2	7.2	15.1	22.3	5	20.8	-	8.7	-	-	3.6	100
Mongolia 2002	64	16	-	0	34	0	12.5	-	19	-	-	-	81
China 1999	1131[c]	25.5	9	0	18	0	24.9	-	13.6	-	-	-	91
China 2002	356	15.2	-	-	27.5	-	22.5	-	14.9	-	-	-	80
Japan 1985	67	7.5	7.5	0	24	3	36	3	9	3	3	4.47	100
LATIN AMERICA													
Chile (g) 1994	79	3.8	5	2.5	8.9	2.5	67.1	-	0	10.2	-	-	100

Table 17.4 Comparison of percentages of anatomical site worldwide

	Cohort	Globe	Glaucoma	Cornea	Lens	Uvea	Retina	Albinism	O.N	ONL	GAN	UD [a]	Total
Chile	318	10.9	7.5	6	9	3	47.5	0	12.7	0	3.4	-	100
Chile 1993	217	8.8	8.3	11	9.2	2.3	47	-	12	0	1.8	-	100
Latin America	-	0	10	8	20	1	26	-	10	25	0	-	100
Ecuador (g) 1995	55	7.2	3.6	0	12.7	0	61.8	3.6	0	0	0	11.1	100
Uruguay	220	22	9	1	25	0	33	-	12	-	-	-	100
Peru 1990	202	15	12	18	12	0	25	-	8	-	-	-	90
Bolivia 1988	78	13	10	23	21	0	23	-	10	-	-	-	100
Jamaica 1986	108	8	15	5	39	0	15	-	18	-	-	-	100
Dominican Republic 1995	51	15	18	18	31	0	10	-	8	-	-	-	100
Argentina	573	24	6	1	8	0	51	-	10	-	-	-	100
Colombia (g)	94	9.5	0	0	14.3	9.5	57.2	-	0	9.5	0	0	100
SUB-SAHARAN AFRICA													
East Africa	491	0	1	72	6	2	3	-	6	10	-	0	100
Ethiopia	295	4.7	1.7	62.4	9.2	8.8	2.4	-	9.8		-	1	100
West Africa	284	8.5	13	36	16	1.1	20	-	5.6	0	-	0	100
Kenya/Uganda	160	12	2	0	10	0	65	-	0	12	-	0	101
Kenya/Uganda (g)	52	11.5	2	0	9.6	0	49.7	15.5	0	0	-	11.6	100
Malawi (g)	24	4.2	0	0	33.3	4.2	50	4.2	0	0	-	4.1	100
Malawi	270	1.5	0	75.2	2.6	0	0.7	0	3	0	-	17	100
Nigeria (2m-14yrs)	140	3	14	21	19	5	11	*	14	4	9	-	100

Table 17.4 Comparison of percentages of anatomical site worldwide

	Cohort	Globe	Glaucoma	Cornea	Lens	Uvea	Retina	Albinism	O.N	ONL	GAN	UD [a]	Total
Nigeria	142	21.4	9.3	21.4	31.4	1.4	7.9	-	7.2	-	-	-	100
South Africa	564	6.4	6.7	11.2	3.7	5.9	38.5	0	15.2	-	12.4	0	100
EME (EUROPE, N. AMERICA)													
Europe & North America	1806	0	2	1	8	1	30	-	23	35	0	0	100
Czech Republic	229	10.9	0	1.8	8.7	5.2	54.2	-	15.3	0	3.9	0	100
Iceland (g)	14	14	0	0	14	0	0	50	0	0	0	22	100
Denmark	150	12.7	0.67	0	6	2.7	18.67	2	27.3	30	-	-	100
Denmark (g)	44	2	2	0	7	2	48	7	0	32	0	0	100
Edinburgh	107	11	2	1	0	0	36	0	50	0	0	0	100
UK													
SVI/BL isolated	102	9.8	6.9	2.9	6.9	5.9	60.8	-	16.7	5.9	-	2.9	119
SVI/BL plus	336	5.7	1.8	1.2	4.1	1.8	19.1	-	31.3	60.1	-	1.5	127
All UK	439	6.6	3	1.6	4.8	2	28	-	28.7	47.8	-	0	123

Figures except cohort size in percentages. ON: optic nerve. ONL: others not listed. GAN: globe appears normal.
* Glaucoma/ albinism cases are also included in the figures for whole globe/albinism but have been excluded from the total.
[c] Figures based on the percentage of the total (1245) examined who fulfilled the criteria of being blind or severely impaired
(g) Genetic Series.

17.8 Aetiologies

Childhood visual disorders can be either hereditary in origin, as a result of chromosomal or genetic defects, or can result from factors operating from the time of gestation throughout the intrauterine period, during birth and the immediate perinatal period, and throughout childhood. (Table 17.4)

A range of clinical entities can result depending on the period of exposure to the ocular insult and the predisposing factors. It is, therefore, possible to divide these non-hereditary factors into; (1) prenatal factors that operate during gestation; (2) perinatal and neonatal factors that operate at the time of delivery (birth) and in the immediate neonatal period i.e. the first month of birth; and (3) post-natal factors operating in childhood. Geographical factors can determine the prevalence of these causes, hence, the variation in different parts of the world. [52]

A precise diagnosis could not be established in all cases. In 60 cases (9%) it was not possible to ascertain the aetiology. Based on the pattern of the clinical conditions in this group, a speculation was which estimates that 80% of these cases are genetic in aetiology which is higher than the percentage of the genetic conditions in the series (77%). It is possible that the higher number of CC in the undetermined group has skewed these results.

17.9 Non-Hereditary Conditions

Prevalence

The non-hereditary conditions on the other hand are among the lowest reported figures, especially in the GS (5.5%), which is less than half that of the WB (12.1%). The average in both regions (12.1%) is very near the Saudi figures for the >1962 cohort although the GS figure is much lower at 5.5%. All the results on non-hereditary conditions in this study are lower than those reported in other Arab countries including Jordan, with striking differences between the high proportion of the pre 1962 Saudi cohort to those of similar ages (25+) where they are less than half those of Saudi Arabia for the same age cohort (Table 17.5).

The low figure for the non-hereditary group is caused by the absence of nutritional factors in the OPT and the low incidence of infections in comparison to Africa and parts of Asia on the one hand, and the scarcity of cases of retinopathy of prematurity in comparison to the developed countries on the other hand. The disparity from the rest of the Arab world is not possible to ascertain, although the vast geography of Saudi Arabia with many remote regions which were not very accessible before the economic prosperity may be the main factor. Neither are Jordanian's higher figures easy to explain although the same may apply to some extent in the author's opinion.

Table 17.5 Temporal variation in the percentages of non-hereditary condition in Saudi Arabia, Jordan, Iraq and this study

	Pre 1962 (Cohort)	Post 1962 S/Arabia; Pre 1970 Jordan' Pre 1972 Iraq	Post 1970/72 Jordan/Iraq 1997-2003 Saudi Arabia
S/Arabia 1985	69% (65)	11.4% (105)	53% (260) [715]
Jordan 1990	-	33% (157)	23.3% (103)
Iraq 1992	-	47.7% (21)	8.5% (9)
This study Jalili 1985-1987			
West Bank	29.8% (47)	21.8% (133)	12% (214)
Gaza Strip	21.6% (51)	16% (125)	5.5% (164)
Total OPT	25.7% (101)	19% (264)	10% (405)

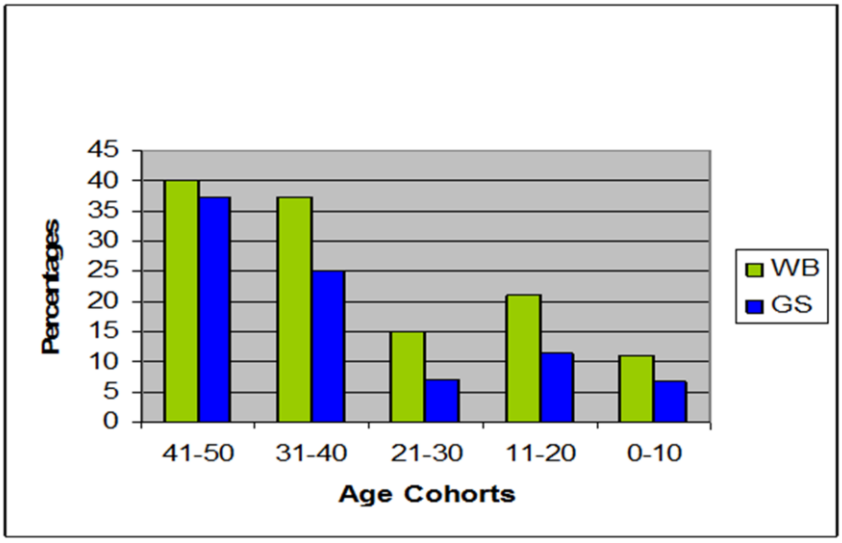
[a] Based on outpatients seen in a referral centre in Saudi Arabia.

Changing patterns

Although the ages in column 'post 62 and pre 70/72' in Table 17.5 are not identical, they show a significant disparity between Iraq, Jordan and Saudi Arabia and reveal the severity of the problem in Iraq before 1972 and the dramatic decline in the non-hereditary conditions in the 1970s, significantly more than that witnessed in Jordan in the same period.

The decline in the non-hereditary conditions in the OPT has been gradual over several decades, although the turning point appears to be in 1952. depicts these changes although data from patients aged 51 years and over were removed as they were exceptionally high (250% and 500% in the WB and GS respectively).

Figure 17.2 Percentage of non-hereditary conditions by region



The remoteness of villages and poor road systems in some of the northern and southern parts of the WB, which is also a problem in the GS, was suspected to cause a higher rate of non-hereditary conditions. Surprisingly, the reverse is true; the percentages of the non-hereditary conditions in these regions are lower than in central regions when compared to the population ratios in these regions which apply to all ages. This cannot be explained on the basis of under representation in these regions and the same argument cannot, therefore, be applied as an explanation for the preponderance of prenatal conditions in the WB (ratio of 3.4:1). This disparity is most significant in the 16+ age group, and becomes less marked in the < 16 and is a reflection of the improved local services in recent years in various parts of the WB such as in Nablus in the north and Hebron in the south of the region, together with the proliferation of various clinics run by charities in the region.

There is a significant drop in the non-hereditary conditions in the < 16 age cohort, predominantly corneal pathology as noted earlier, which clearly demonstrates a drop in the percentage of anterior segment infections over the years. In the WB the percentage drops from 15.8% to 4.7% and in the GS from 15.2% to 1.2%. (Figure 17.2) The acquired conditions formed 87% (n= 16) of patients above the age of 16 years, and 77% of the total corneal category.

This finding is in keeping with other studies in the Middle East as outlined in Chapter 9.

No disparity exists in the prevalence of the aetiological groups between villagers, town and camp residents in this study contrary to the findings in Egypt where 2/3 of the blind had a rural residence.

Although trauma is not a major contributor to blindness in this study, it is an important cause of unilateral blindness based on the author's observations and the lack of playground facilities is an important factor.

Comparison to Worldwide Data

The WB and the GS figures on non-hereditary conditions fall in the lower bands of worldwide When the is compared to figures from the rest of the world, the following patterns are demonstrated: -

Non-hereditary conditions from childhood causes

The WB and the GS data between 1985 to 1987 fell in the lower bands of worldwide prevalence of non-hereditary conditions.

- <10% band: Gaza Strip with its 5.4% non-hereditary conditions in the < 16 falls into the same band as the combined WB and GS data in 1992, [27] Iraqi post 1972, [17] Uzbekistan, [6] and Sri Lanka. [53], [54]

- 10-20% band: The WB with its 15.5% ratio is in the same rank as Saudi Arabia post 1962, Lebanon in 1975, [7] Cyprus, [40] and Mongolia. [55]

- 20-30% band: Neighbouring Jordan post 1970 falls in this band together with north Israel in 1992, [5], [56] South Africa, Kenya and Uganda, [35], [48], [57], and Malaysia. [58] India's figures are slightly higher averaging 31.5%. [18], [36]

- Higher bands with >40% non-hereditary conditions include the Philippines, Thailand, [59] most of the African countries including Nigeria, [24] [49], Ghana, Togo, Benin, [18] with the highest figures in Malawi at 62%. [45]

Non-hereditary conditions predominantly from intra-uterine causes

The prevalence rates vary with Jamaica [60] and Peru [61] with a 40% prevalence of IU causes; the Nordic countries with 60% and Ireland with 80% non-hereditary conditions, half of them are the result of intrauterine factors. [28]

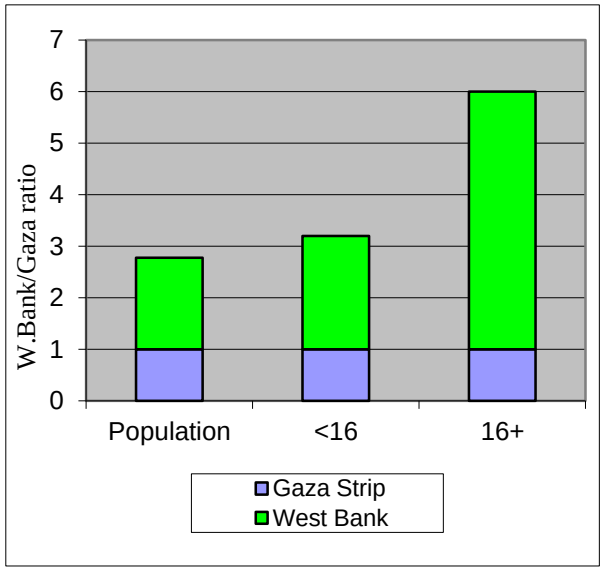
Non-hereditary conditions predominantly from perinatal causes

Perinatal causes are the main aetiology in developed countries and it is primarily as a result of prematurity. [42] In the middle-income countries such as Czech Republic and Chile, ROP is appearing as the major cause of blindness and forms the highest subcategory of the non-hereditary conditions. [9], [21]

Prenatal Conditions

Prenatal conditions form an insignificant group of 24 cases (3.4% of the series) and the perinatal are even lower at a mere 10 cases (1.4% of the total series). Figure 17.3 demonstrates the decline in this aetiological group in the < 16 age group in the study.

Figure 17.3 Prenatal conditions - comparison between WB :GS population ratio



The majority of prenatal conditions were undetermined IU infections as seen in (Chapter 15, Table A15.1). They share in common their high rate of miscarriages, where the ratio of miscarriages per family is 1.3:1 compared to 0.5:1 and 0.7:1 in the perinatal and postnatal conditions respectively. The exact nature of the causative IUI is undetermined, however, the

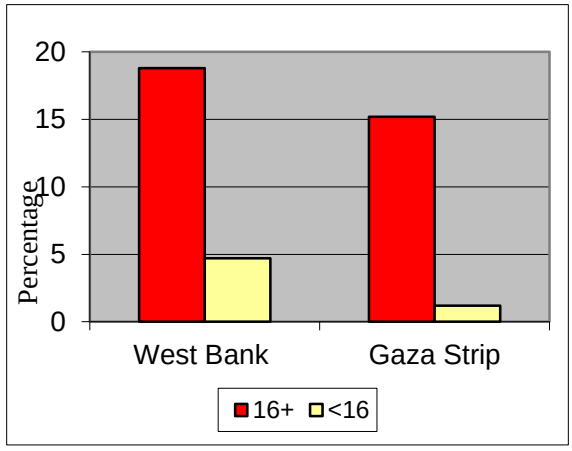
presence of multiple miscarriages in successive pregnancies is highly suggestive of CMV infections. [28], [39], [62]

Perinatal Conditions

Perinatal conditions are an insignificant group made up of 10 patients; 4 were ROP (all dealt with in Israel, and the remainder were due to, birth hypoxia, ophthalmia neonatorum, iatrogenic causes, traditional medicine and surgical complications. (Chapter 15). The low percentage of perinatal conditions in the study (2%) contrasts to the higher levels found in Northern Israel [44], middle income countries [97, 121] and the technically developed countries [90, 95] for reasons mentioned earlier but it equates with those in the rest of the developed world including the Middle East [23, 33, 86, 88]. The number is insignificant in comparison with the magnitude of the problem in other developing countries especially SSA where they form the bulk of the causation of blindness in children. [18], [45] This decline is due to the combination of improved health care, better nutrition and the availability of ophthalmic services at a relatively close proximity. The only exception is Sudan, in particular southern Sudan, where acquired anterior segment conditions from VAD, measles and trauma remain the primary cause of blindness accounting for 40% of cases. [64]

The second group of conditions in the childhood onset conditions are the neurological subcategory; mainly the result of CNS pathology which encompass meningitis/encephalitis, intracranial tumours, intracranial thrombosis, spontaneous subdural haemorrhage, cortical blindness and head injury. These, excluding the trauma, form 23.6% of the postnatal causes. The pathology of CNS causes in this study differed from that found in developed countries where they are predominantly from the effect of prematurity. [28], [42], [43], [62], It is interesting to note that these conditions contribute 19% of the causes in Mongolia with meningococcal meningitis being a predominant preventable cause of SVI/BL whilst in this study the group

Figure 17.4 Changing trends in acquired blindness by region



forms only 2% of the total causes with meningitis/encephalitis less than 0.9%. [37]

Postnatal (Childhood) Conditions

The largest group in this subcategory is ocular infections constituting 36% of these conditions. The number is insignificant in comparison with the magnitude of the problem in other developing countries especially Sub Saharan Africa (SSA) where they form the bulk of the causation of blindness in children [18], [45] (Table 17.4)

The second group of conditions in the childhood onset series are the neurological sub-category; mainly the result of CNS pathology which encompasses meningitis / encephalitis, intracranial tumours, intracranial thrombosis, spontaneous subdural haemorrhage, cortical blindness and head injury. These, excluding trauma, form 23.6% of the postnatal causes. The pathology of CNS causes in this study differs from those found in developed countries where they are predominantly from the effect of prematurity. [28], [42], [43] [62], It is interesting to note that postnatal conditions contribute to 19% of the causes in Mongolia with meningococcal meningitis being a predominant preventable cause of SVI/BL whilst in this study the group

forms only 2% of the total causes with meningitis/encephalitis less than 0.9%. [37]

The third group, trauma, forms 20% of the postnatal category affecting 11 cases in total, including 1 case of head injury, (1.6% of the series) with a slightly higher prevalence in the GS (ratio 1.2:1). Male preponderance was not marked at only 1.2:1. Apart from the single case caused by head injury, the remainder were direct ocular injuries, including chemical. Trauma as a cause of blindness escalated in later years and injuries rose sharply, a staggering 32 and 9-fold increases in the first and second uprising 'Intifida' respectively. Before this, victims were men and there were no <10's. During the Intifida 9% were women and 12% were under 14 years. Injury patterns changed with 65% due to firearms or explosives, 19% to beating and 6% to gaseous substances. Among children, most firearm injuries involved the head, including eye injuries and brain damage. [65] These injuries are usually the result of direct assault by the military and differ from parts of the world where they are caused by landmines such as Cambodia and Afghanistan. [66], [67]

17.10 Hereditary Conditions and the Role of Consanguinity

In industrialised countries, hereditary conditions are an important cause of childhood blindness, accounting for 16-51% of cases. [44] This applies to our findings in both Palestinian regions although the proportion is higher in the GS (84.4%) than the WB (76.4%). This disparity evens out when the number of pedigrees is included (WB 65.6%, GS 64.3%), thus indicating that the true incidence of genetic mutation is equitable in the two regions.

In this study, heredity was the largest aetiological group with 519 patients from 310 sibships (243 pedigrees). Non-hereditary conditions formed a smaller group comprising 90 patients and sibships with 89 pedigrees. In 60 patients aetiology was undetermined.

The percentages of hereditary conditions in the OPT are among the highest recorded in world literature and the GS figure of 86% in the < 16 ranks the top. This is a result of the very high rate of consanguinity in the OPT which, after painstaking study in tracing the consanguinity, appears to be higher in the GS. The OPT figures compare well with findings from other countries in the region. [5] [6], [7], [20], [40], Indeed the WB average figure of 76.4% is almost identical to Jordan's post 1972 figure (78%), [5] and the GS average of 84.4% is close to the figures from Saudi Arabia post 1962 (80%). [20] It is interesting to note that the large GS pedigree with multiple sibships and a large number of patients originated some centuries earlier from Mecca, Hijaz in the Western region of Saudi Arabia. [68] On the other hand, Palestinians in the WB have always had closer links to those in Jordan. The geographical proximity of the WB to Jordan, and the GS to Egypt, together with the separation of the two regions by Israel, whether before the 1967 war or after the occupation, with the resultant frequent road blocks, have enhanced the isolation of the two regions in the four decades that preceded this survey.

. It is worth noting that figures for the Palestinian population in north Israel is less than half of their counterpart in the OPT at 38.3% and lower figures for genetic diseases were also reported in Uzbekistan, another Muslim country with high rates of consanguinity. [6], [69], [70] In the former, it was postulated that an ethnic genetic factor existed causing an increased risk of genetically determined conditions in children of Muslim communities (66%) as compared to other communities of Druze and Christian (14% to 42%), despite the fact that consanguinity was higher in the Druze (85%) and Christian (77%) groups respectively.

The proportion of Christians in this survey is too small to draw any solid conclusions from, although the author of this study has observed from the cases he encountered throughout his two year practise at SJOH, that retinal dystrophies among Christians tended to be of a teenage onset and took the form of the recognised clinical types of RP described in the

West. The predominance of CNS conditions in Christians is difficult to explain although the sample size is too small (n=5).

The figures from the Arab World are not unique; they share the high percentage of hereditary conditions with neighbouring Cyprus (79%), [6] [40], a predominantly Greek Orthodox country where first cousin marriage is not practised but the population has become genetically isolated because of its small size and population. In comparison to other Muslim countries, figures from Arab countries are higher than those from Uzbekistan (54.5%) and Turkey (25%). [6], [71] In the UK, the incidence of genetic disorders in the Pakistani population is 10-fold higher than in other ethnic group. [72] In the latter, however, a high percentage of cases of unknown aetiology were reported (45.2%), some of which could have been genetic in origin.

Consanguinity

Consanguinity, particularly between close relatives, is known to increase the risk of recessively inherited disease and multifactorial disorders, but the effects of inbreeding on the prevalence and type of AR disease in a community are complex and difficult to quantify. [73] Many different factors are involved, ie the degree of relationship of the parents; the number of generations over which inbreeding has been practised; whether the genetic abnormality alters biological fitness and the mutation rate of the genetic abnormality. [74]

In this study two figures have been highlighted; those of the hereditary conditions and those of the non-genetic conditions. The former reveals a high rate of consanguinity among the sibships in the hereditary conditions category (see 17.10) (Table 17.8) These agree with the common finding of increased morbidity in association with consanguinity. The trend of consanguineous marriage in the hereditary series has shown an increase over the decades climbing from 67.6% in sibships of patients over 30 years, to 74.4% in the under 16 cohort; a trend which is more noticeable in the GS, rising from 68.2% to 85.2% in the corresponding cohorts. This is attributed to increasing social isolation as a consequence of the ongoing unrest, together with an increase in the isolation of the remaining affected sibships, caused by local knowledge of an existing condition compounding the problem further.

The figures for consanguinity drop in the non-genetic cohorts to 72% in the WB and 74% in the GS; but this decrease widens in the < 16 cohort to 47% in the WB, and 66.6% in the GS. This suggests a decline in consanguinity in the general population, mainly in the WB, whose figures in general resemble those of Jordan ranging between 49.3% and 58%.

First cousin marriages are well documented as the commonest type of marriages in this highly inbred population and this is reflected in the study where the rate of hereditary (recessive) disorders is the highest recorded among the studies. (Table 17.7) The author believes this degree of accuracy is due to the detailed histories taken and pedigree charts drawn as part of the study, reducing the rate of undetermined cases and enabling more accurate results. In addition there is a predominance of conditions known to be inherited in an AR manner. These potentially large pedigrees provided the opportunity for gene tracking, gene mapping, and the investigation of phenotype variability.

In the MEC the high proportion of genetic disease due to AR conditions (50-65%) has also been caused by the high levels of consanguineous marriages which range from 16-55% of all marriages. [75]

In other countries that are known to practise consanguineous marriage (Sri Lanka and India), the proportion of children with visual loss from AR disease was also high (52%). In Kenya and Uganda, 44% of children with genetic disease had AR disorders, which is partly explained by the relatively high proportion of children with oculocutaneous albinism (15%). In Ecuador, a largely Catholic country, 54% of children with genetic disease had recessively inherited disorders.

17.11 Changing Patterns

Our findings indicate a shift of causation over the decades from anterior segments and infective disorders, which were prevalent at the beginning of the last century to posterior segment and hereditary conditions which characterise developed countries. This is despite the poverty prevailing in large parts of the region. The prevalence of childhood blindness (CB) has come down from 154/100,000 at the beginning of the last century to 20/100,000. In comparison, the prevalence of CB in Sudan is 140/100,000 which compares to the old Palestinian figures. **(76)**, **(77)**, **(64)**

The study has also shown that the commonest cause of childhood blindness in the Occupied Palestinian Territories (OPT) is genetic and the severity of the blindness varied with the condition and its duration. The visual morbidity was very severe from the start such as in Leber congenital amaurosis (LCA), microphthalmia (MC), congenital glaucoma (CG), or progressive such as in cone and cone rod dystrophies (Figure 19.6). Ranking at the top of the league of 104 worse visual acuities (Category '5', NLP) are whole globe conditions (55.8%) with genetic conditions occupying two thirds of the whole globe (anophthalmia/microphthalmia 31%, CG 26%) and postnatal causes one-third in the form of phthisis (27.5%). This is very much in line with the patterns of blindness seen in some affluent Arab countries where there has been a shift from anterior segment pathology from malnutrition and infections to genetic causes. **(64)**

Health services in the region and premature babies care is lacking at the time of the survey, hence the near absences of causes resulting from prematurity seen in the middle-income and established market economy countries in the form of retinopathy of prematurity (ROP) in the former and central causes in the latter. The OPT figures compare with the wealthier Arab countries and differ from those seen in the poorer countries such as Yemen and Sudan where onchocerciasis and vitamin A deficiency (VAD) are the predominant causes. This also applies to optic nerve disorders which are a significant contributor to childhood blindness in Established Market Economies (EME).

Prevalence of Blindness and Visual Impairment

The prevalence of childhood onset blindness is worked out on the basis of average population figures in each region in the period of the study (section 3). Numbers of patients in the relevant categories are found in Table 16-1.
Prevalence of SVI/BL and blindness per 100,000 is at least as follows:

Table 17.6 Prevalence of blindness and visual impairment

	per 1000 Population		
	West Bank	Gaza Strip	Total
SVI/Blindness			
<16	28	28	30
All ages	24	30	27
Blindness			
<16	19	9	21
All ages	18	22	

17.12 Avoidable Blindness

Avoidable blindness in the OPT has dropped in the <16 age group, significantly more in the GS where it is half that of the older cohorts; this has been addressed earlier. (Table 17.8)

The preventable causes in the WB compare with those in South India, which falls between West Africa and Chile. The problem appears to be considerably lower than some of the neighbouring Arab and Muslim countries.

In Egypt for example more than 50% of childhood blindness is preventable and in Turkey the proportion is even higher where avoidable causes accounts for as much as 70% of causes.

Congenital cataract is the main treatable condition whilst the severity of the CG makes

blindness almost inevitable in these conditions; prevention would be through tackling consanguinity in this community. This also applies to the rest of the hereditary conditions which are potentially preventable as the majority are recessive from high inbreeding and a long-term campaign to combat consanguineous marriage should help in reducing the prevalence of these conditions.

The disparity in the WB in cataract cases in the study between children and adults is probably due the combination of a lower representation of adults in the WB cohorts on one hand and an increased awareness of the need for surgery in younger generations on the other hand. This demonstrates a true predilection of the conditions to females. The same can be applied to RCD.

17.13 Recommendations

As the bulk of the conditions are AR hereditary conditions as the result of the high inbreeding, these are theoretically preventable by reducing this practice. This, however, is very difficult because of tradition, lack of awareness of the harmful effects of this practice and other socio-economic and political factors. A long-term educational programme is necessary which needs to be addressed at all levels of the community and conducted by all groups concerned; including the authorities, educational bodies, charitable organisations and also, most importantly, religious bodies whose preaching on this subject is essential.

The other recommendation is the need for continuity of care and supervision of common conditions such as CC and CG. Cataract patients are left without aphakic glasses until they are school age, leading to intractable amblyopia. Frequently glasses are not prescribed after surgery because of the knowledge that they will not be used. This explains why nearly 45% of these patients are SVI/BL. Congenital cataract is the commonest treatable condition in the region. A mechanism is needed to monitor this problem which should include supervision at home, and financial support to provide spectacle for children and replace them when needed.

Glaucoma is considered a treatable condition in the various literatures; however, the condition in the WB and the GS is very severe, with a poor prognosis. A large proportion of technically successful surgeries, including corneal decompensation, fail. Treatment is also neglected in those who require long-term treatment with drops caused by financial constraints.

Towards the end of the study, a proposal for a programme for the prevention of blindness in the region was put forward by the author and was adopted by the Christian Blind Mission (CBM) and its supported charity, the Friends of the Patient, in Bethlehem. Funding was allocated to the project but unfortunately it had to be suspended because of the civil unrest that started at the end of 1987 and is still continuing. [78]

Genetic counselling is another required measure that is lacking (and to my knowledge is still lacking). Prenatal genetic testing may be rejected in some sectors of the community on religious grounds. Premarital carrier matching, which was found acceptable among Muslim Bedouin communities in Israel, [79] may be an alternative method to reduce genetic disorders.

Specialised paediatric ophthalmic services in the WB (SJEH) and the GS would be a step forward in the proper management of the frequent paediatric problems.

Table 17.7 Proportions of Hereditary and Non-Hereditary Causes in World Literature

Country	Cohort	Hereditary	Total NH	Intrauterine	Perinatal	Childhood	UD
Total Study < 16	405	***76.5***	***15.5***	***4.6***	***2***	***8.9***	***8.2***
16+	264	***79.2***	***19***	***2.3***	***1.1***	***16***	***0.7***
Total Series	669	***77.6***	***13.4***	***3.7***	***1.5***	***8.2***	***9***
WB < 16	214	***76.2***	***11.6***	***5.1***	***1.9***	***4.7***	***7.2***
16+	133	***76.7***	***21.9***	***3.8***	***2.3***	***15.8***	***0.6***
WB Total	347	***76.4***	***15.5***	***4.6***	***2***	***8.9***	***7.8***
GS < 16	164	***86***	***5.4***	***2.4***	***1.8***	***1.2***	***4.8***
16+	125	***82.4***	***16***	***0.8***	***0***	***15***	***0.7***
GS Total	289	***84.4***	***10***	***1.7***	***1***	***7.3***	***5.5***
ME and MEC							
Jordan 1992 <1970	157	***67***	***33***	***4***	***4***	***25***	-
>1970	103	***78***	***22***	***4***	***7***	***11***	-
Saudi Arabia 1985 <1962	65	***25***	***69***	***3***	***0***	***66***	-
>1962	105	***84***	***13***	***1***	***1***	***11***	-
1997 - 2003	273	***53***	***47***	-	-	-	-
Lebanon 1975	203	***77***	***23***	***1***	***2***	***20***	-
WB and GS 1993	173	***55***	***8***	-	-	-	***37***
Egypt	113	***48.7***	***51.3***	-	-	-	-

Sudan 2005		40	***10***	***85***	***-***	***-***	***-***	***5***
Iraq 1990	1961-70	44	***52***	***47.7***	***-***	***-***	***-***	-
	1970 -80	106	***91.5***	***8.5***	***-***	***-***	***-***	
Northern Israel 1992 (infants)		193	***38.3***	***29.6***	***8*** [a]	***24.4***	***0.5***	***32***
Cyprus 1972		112	***79***	***19***	***4***	***2***	***13***	***2***
Uzbekistan 1999		671	***54.5***	***5.4***	***1.4***	***0***	***4***	
ASIA								
India (9 states) 1995		1318	***23***	***31***	***1.8***	***1.4***	***27.9***	***46***
India (average) 2001		2211	***26***	***32***	***2***	***2***	***28***	***42***
North India 2003		703	***13.4***	-	***-***	***-***	***28***	***56.5***
Sri Lanka 1995		226	***35***	***8.8***	***-***	***-***	***-***	***56.2***
Thailand/the Philippines 1993		244	***16.8***	***45.5***	***5***	***1***	***66***	***37.7***
Khoa-Kaen (Thailand)		65	***13.8***	***35.6***	***3.1***	***20***	***12.5***	***50.8***
Manila (Philippines)		113	***17.7***	***40.7***	***2.7***	***23***	***15***	***41.6***
Baguio (Philippines)		31	***9.7***	***61.3***	***0***	***0***	***61.3***	***29***
Davao (Philippines)		35	***17.1***	***59.1***	***0***	***2***	***57.1***	***22.9***
Malaysia 2001		332	***29.5***	***21.3***	***4.5***	***9***	***7.8***	***49.1***
Mongolia 2002		64	***27***	***17***	***-***	***-***	***-***	***48***
China 1999		1131	***30.7***	-	***-***	***-***	***14***	***52.9***
AFRICA								
Ghana, Togo, Benin 1993		284	***21.1***	***45***	***-***	***-***	***-***	***34.9***
Kenya & Uganda 1995		160	***32.5***	***28.1***	***-***	***-***	***-***	***39.4***
Uganda 1998		1135						***43.6***

(School age)	-	***12.9***	-	***0.5***	***0.5***	***42.7***	***43.6***
(Community)	-	***12.7***	-	***3.5***	***0.7***	***33***	***50***
Nigeria (2m-14yrs) 1970	140	***25***	***57***	***7***	***1.4***	***91.7***	***18***
Nigeria 2003							
8-15	46	***19.6***	-	***4.3***	-	***37***	***39.1***
>15	94	***12.7***	-	***9.6***	-	***39.4***	***38.3***
Malawi (1976)	218	***11***	***62.4***	-	-	-	***26.6***
South Africa 1997	564	***33***	***25.5***	***0.9***	***13.1***	***11.5***	***41.5***
LATIN AMERICA							
Chile 1994	264	***29.6***	***41.9***	-	-	-	***28.5***
Chile							
<10	141	***24.9***	***41.9***	***10.6***	***26.9***	***4.3***	***33.3***
11+	126	***34.1***	***42.1***	***5.6***	***17.5***	***19***	***23***
Colombia 1995	94	***22.4***	***34***	-	-	-	***43.6***
Ecuador 1995	142	***38.7***	***35.9***	-	-	-	***25.4***
Jamaica 1988	108	***48***	***52***	***41***	***0***	***11***	***0***
Peru 1990	202	***51.1***	***43.5***	***41***	-	-	***5.4***
EUROPE							
England & Wales 1967	776	***50***	***50***	***6***	***33***	***11***	-
England & Wales 1996 (0-15)	728	***46.5***	***52.5***	-	-	-	***1***
UK (All) 2003	439	***32.8***	***64.1***	***28.8***	***17.5***	***17.8***	***9.1***
Edinburgh 1987	99	***48***	***52***	-	-	-	-
Edinburgh 2002	107	***26***	***48***	***3***	***40***	***5***	***26***
Northern Ireland 1977	486	***51***	***27***	-	-	***5***	-

Ireland 1991	172	***16***	***34***	***40***	***27***	***13***	***40***
Denmark 1987	150	***29.3*** [b]	***55.9***	***20.6***	***16***	***19.3***	***14.6***
The Nordic Countries 1992	2527	***35***	***33***	***-***	***-***	***-***	***32***
The Nordic Countries 1996	304	***5.6***	***94.6***	***64.5***	***20.7***	***9.2***	-
Netherlands 1996	1300	***45***	***26***	***-***	***-***	***-***	***29***
Czech Republic 2001	229	***9.2***	***48.5***	***0.4***	***43.7***	***4.4***	***42***
Australia 1968	44	***50***	***50***	***20***	***16***	***14***	-

[a] Infections not specified whether intrauterine or postnatal [b] Percentages extracted by the Author of this thesis.
UD: Undetermined
Percentages in ***bold Italic***

17.14 Regional Differences

The two Palestinian regions have certain distinguishing features that have been intensified in recent decades as a result of the physical separation after 1948 by the formation of the State of Israel and the annexation of the WB to the Kingdom of Jordan and the GS to the Egyptian administration. This geographical separation lessened after the 1967 war and the occupation of both regions by Israel. This enabled some contact via Israel. However, for both military and practical reasons, this was not always easy and was frequently interrupted by roadblocks and the frequent closure of borders. School curricula, healthcare and economy followed the Egyptian system. The WB on the other hand was an integral part of the Jordanian system as were the movement of people and contact. This survey has demonstrated certain difference in the pattern of visual impairment, disease and prevalence between the two Palestinian regions.

Demographical Aspects

The main differences lay in the higher number of patients per population the GS which has resulted from larger number of patients per sibship. The affected pedigrees ratio between the two regions (mutation rate) remains marginally higher in the GS but close to the general population ratio.

Hereditary conditions are also higher in the GS but the non-hereditary cases are significantly lower in the GS, especially in the young and school age pupils, thus demonstrating a significant decline in these conditions in comparison to the WB at the time of the study. Consanguinity rates are also higher in the GS with a notable difference in that there is an increase in the consanguinity rate among the younger generations under the age of 30 years in comparison to the WB where the rate has remained steady over the years. Autosomal recessive (AR) conditions are also predominant in the GS although autosomal dominant (AD) traits barely exist in the WB. (Table 21.2)

Gender Difference

Demonstrable differences exist between the WB and GS in the M:F ratio in several conditions and similarities in others. Table 21.3 depicts the conditions according to their male and female preponderance or equal gender in both regions. The general trend is a male preponderance in the GS and female preponderance in the WB. Several conditions show a significant WB predominance; most significant of all are rod disorders. Other smaller cohorts also show the same trend. On the other hand, the GS has its own characteristics with the preponderance of all the cone disorders especially the CRD and achromatopsia, in addition to smaller categories such as hypopigmentation, microphthalmia/anophthalmia, myopia and congenital idiopathic nystagmus. The latter is caused by the presence of one extended pedigree with the condition.

The total series of RD is the only group of conditions that demonstrate a ratio equitable to that of the general population. Congenital cataract comes next with a WB:GS ratio close to the population ratio in the total cohort but with GS predominance the school age cohorts.

Several conditions show a significant WB preponderance; most significant of all is the large series of rod disorders. Other smaller cohorts also show the same trend. On the other hand, the GS has its own characteristics with the preponderance of all the cone disorders especially the CRD and achromatopsia, in addition to smaller categories such as hypopigmentation, microphthalmia/anophthalmia, myopia and congenital idiopathic nystagmus. The latter is caused by the one extended pedigree with the condition.

Table 17.8 Comparison between the WB and GS: population, patients, conditions inheritance and blindness, 1985-1987.

Parameter	WB	GS
Demography / population		
OPT[a] population in 1000s	1091	604
School age	562.3	322
WB:GS ratio	1.78:1	
school children	1.75:1	
The Survey		
M:F Ratios (total series)	1.1	1.6
(School age)	0.96	1.56
Hereditary [b] (school Age)	0.87	1.25
Non-Hereditary	3.75	6
Hereditary -WB:GS	1.3	
Non-hered. WB:GS	2.7	
Affected cases	347	289
WB:GS Ratio	1.2	
Affected sibships	256	170
WB:GS Ratio	1.5	
Affected Pedigrees	228	131
WB:GS Ratio	1.74	
Sibship:Pedigrees ratio	1.2:1	1.3:1
Hereditary conditions		
<16	76.2%	86%
16+	76.7%	82.4%
All ages	76.4%	84.4%
Genetic retinal conditions	96%	99%
Non-hereditary conditions		
<16	12%	5.5%
16+	22%	16%
All ages	16%	10%
Consanguinity		
% Hereditary (<16, 16+)	82%	95%, 90%
% Non-hereditary (<16, 16+)	72%	78%, 71%
Consanguinity per 15 years of age: <15, 15-30, 30+	65%, 64%, 56%	85%, 80%, 68%
Mode of Inheritance in the hereditary series		
AR	86.4%	92.6%
AD	3.4%	0.8%
Prevalence SVI/BL		
% of SVI/BL		
SVI/BL school age	64% (125)	47% (65)
SVI/BL All series	67% (231)	56% (163)
BL school age	46% (90)	34% (47)
BL All series	51% (176)	41% (119)
Prevalence per 100,000 population		
SVI/BL - School children	24	20
All series	22	27
Blind School children	17	16
All series	16	20

OPT: occupied Palestinian territories
[a] Hereditary Conditions in the total series 1:1

17.15 Overall

It is evident that patterns of blindness are strongly influenced by the levels of wealth and facilities of individual countries and areas together with social customs. In general, as a country develops the wealth and resources to era dicate bacterial causes of blindness, these are replaced by genetically determined causes. The degree of such causes can also vary depending upon the structure of the society, ie whether the population is isolated and thus forms an endogamous group or whether consanguinity exists. As many of the developing countries are still under the influence of blindness acquired through infections and have rapidly expanding populations, we can anticipate that even when causes of acquired blindness are lessened, they will still face a serious problem from genetic disorders in enlarged populations.

Equally, it remains difficult to have an overall picture of the incidence and prevalence of childhood blindness and visual impairment. Whilst more researchers are using comparable protocols and definitions, there is still a lack of

unanimity in the sources and style of research. Various researchers have drawn attention to the shortcomings inherent in either blind school surveys, blind registrations or population based surveys; also coupled with examiner bias. At present, individual surveys are able to make recommendations on the sample size they see, and indeed more information is coming out on regional differences and needs. However, the situation remains whereby the causes and methods of prevention remain very different in developed and developing countries/regions whilst, at the same time, we have seen the problems faced by countries that develop at an accelerated rate.

A protocol for the management of CC in the context of the developing world is being developed at the International Centre for Eye Health. The management of infantile cataract in countries with advanced economies is highly labour-intensive and costly and must be adapted to situations where facilities and finance are not so freely available, if these children are to have the best achievable vision given the limited resources obtainable. [80]

Table 17.9 Gender differences in clinical conditions by region

Condition	WB [a]	GS [a]	WB: GS	Cohort
General Population ratios *n:1*				
< 19 years	*1.1*	*1.08*	-	-
> 19 years	*0.86*	*0.9*	-	-
All ages	*0.88*	*1*	-	-
Male Preponderance				
CC [b]	-	-	*1.8*	132
CG				
Total series	+	-	*1.5*	68
< 6	+	-	*4*	20
Small Eyes	+	*2, 1.4*		15, 22
Optic Nerve	*+, 1.8*	-		23
Myopia	-	+		24
Albinism	-	+	*19*	2.2
All rod disorders	-	+	*1.2*	22
RCD	-	+	*2*	2
Female Preponderance				
CG (6-18)	*+ 0.6*	*+ (0.9)*		WB, 15; GS,14
Achromatopsia	+	*0.3, 0.8*		33
Uvea	+	-	*0.6, 0*	6, 0
RD	+	-	*0.8, 1.2*	144
CD	+	-	*0.6*	8
CRD	+	-	*0.6. 1.2*	8
Albinism	+	-	*0.3*	8
All RCD	+	-	*0.7*	87
LCA	+	-	*0.9*	73
Childhood RP	+	-	*0.3*	17
CIN [c]	-	+	*0, 0.4*	7
Equal sex ratio (M:F ratios between 1.0 to 1.2)				
CD	+		*1*	12
RDes	+		*1*	151
Uvea	+		*1*	6
CRD	+		*1*	47
School Age				
Rod 6-16	+	-	*1*	60
CACR	+	-	*1*	11
Myopia	+	-	*1*	8
MC (6-18)	-	+	*1*	8
LCA	-	+	*1*	19
All cone		+	*1.1*	65

[a] Ratios in *Italic*
[+] Male or female preponderance

Table 17.10 Comparison between the WB and GS: prevalence of clinical conditions. West Bank / Gaza Strip Prevalence of Clinical conditions

	School Age		All Ages		Pedigrees [a]	
	Ratio	Cohort	Ratio	Cohort	Ratio	Cohort
Equal Proportions						
Retinal Dystrophies	**1.7**	129	**1.4**	245	**3**	65
Cong. Cataract	**1.2**	56	**1.6**	120	**1.4**	70
WB Predominance						
Optic Nerve	**11**	12	**4**	23	**6**	21
Vitreo retinopathies	**9**	10	**5.5**	13	**6**	7
All Rod	**5**	60	**4**	112	**5.3**	57
LCA	**5**	50	**4**	94	**4.8**	39
Childhood onset	**1.3**	7	**5.6**	21	**6**	13
CACR	**6/0**	6	**11/0**	11	**6/0**	6
Uvea	**3/0**	4	**8**	6	**5/0**	5
Gaza Strip Predominance						
Small Eyes	**1.1**	17	**0.7**	37	**1.2**	18
Mac. Degenerations	**1**	12	**0.8**	22	**2.6**	11
All cones	**0.6**	49	**0.5**	103	**1.5**	30
CRD	**0.2**	24	**0.2**	47	**1**	10
RM	**0.5**	12	**0.3**	33	**0.8**	11
Cong. Nystagmus	**0.3**	4	**0.16**	7	**0.3**	4
Hypopigmentation	**0.3**	18	**0.4**	27	**0.4**	10
Myopia	**0.3**	12	**0.5**	24	**0.6**	13

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