

16 ANATOMICAL CLASSIFICATIONS

16.1 Introduction

Based on the protocol outlined in the WHO/PBL Eye Examination Record for Children with Blindness and Low Vision, Coding Instruction, conditions are reclassified anatomically and by underlying cause ^{11, 12}. Where there was a difference in the anatomical diagnoses between the two eyes, then diagnosis of the best seeing eye was used. These anatomical sites include the whole globe, cornea, lens, uvea, retina, optic nerve, and others not listed. The anatomical sites of the primary pathologies are shown in Table 16-1.

In this survey, the main anatomical sites of pathology are the retina, the lens, and the whole globe. When glaucoma is analysed separately rather than part of the whole globe, it takes the third place followed by the rest of the whole globe. (Figure 16-2a and 2b). The top 4 anatomical sites for primary pathology are demonstrated in Figure 16 –1.

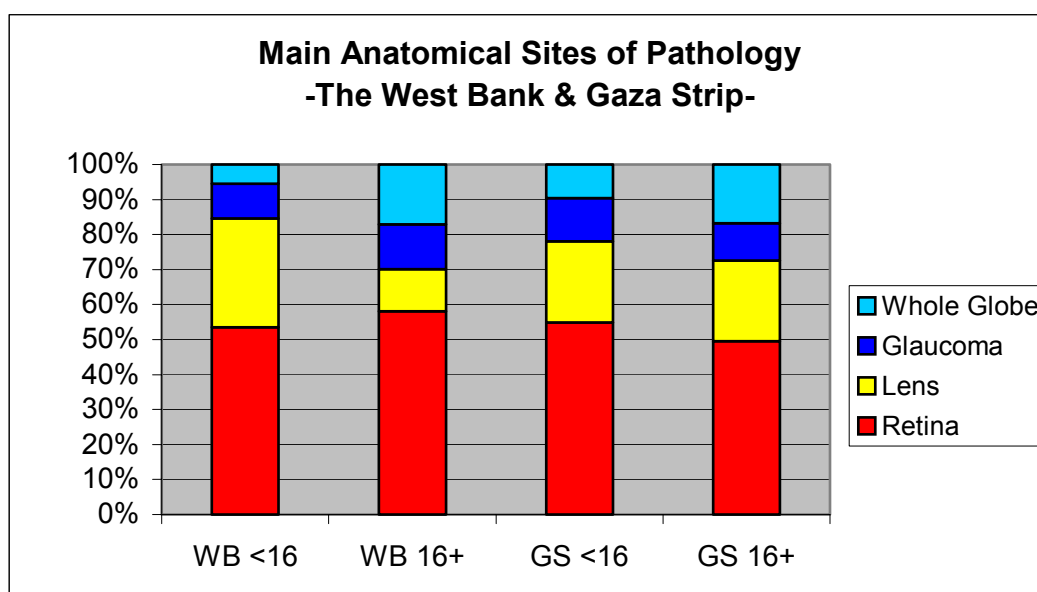


Figure 16-1: Main anatomical sites of pathology by region

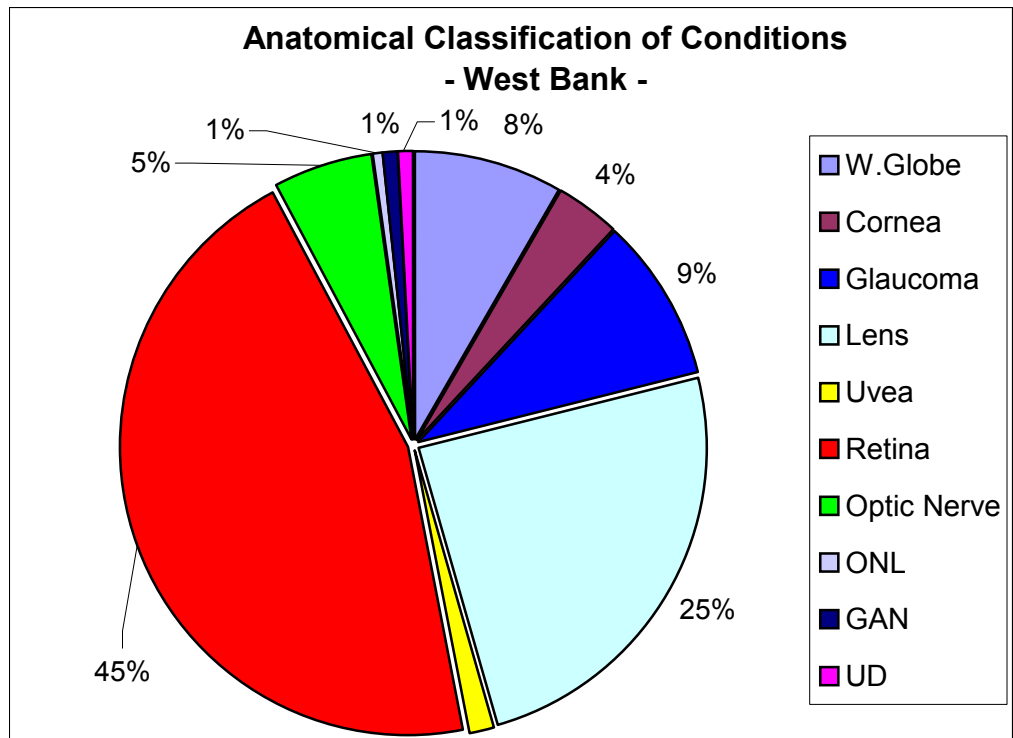


Figure 16-2a: Anatomical classification of conditions in the West Bank

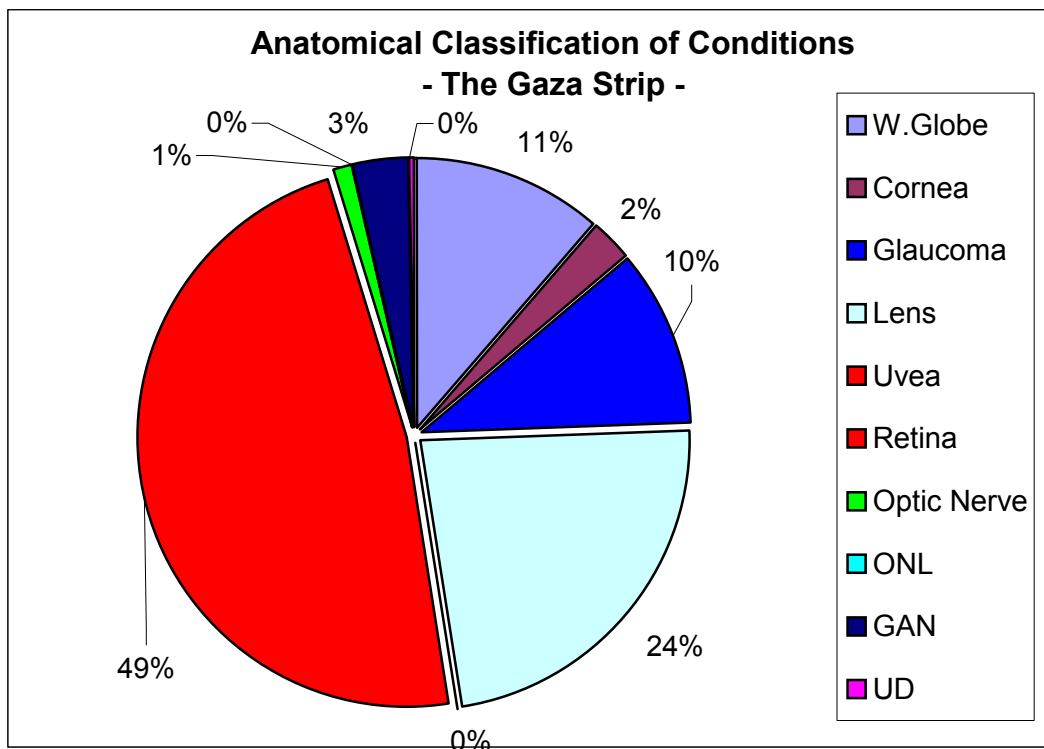


Figure 16-2b: Anatomical classification of conditions in the Gaza Strip

Table 16-1	West Bank						Gaza Strip						All Series						WB/GS Ratio		
Anatomical Site	<16	16+	Total WB			<16	16+	Total GS			<16	16+	All ages			<16	16+	All			
Whole Globe																					
Not specified	0	-	1	0.8	1	0.3	0	-	2	1.6	2	0.7	1	0.2	3	1.1	4	0.6	-	-	-
Phthisis	1	0.5	7	5.3	8	2.3	1	0.6	9	7.2	10	3.5	2	0.5	16	6.1	18	2.7	1.0 :1	0.8 :1	0.8 :1
Anophthalmos	1	0.5	1	0.8	2	0.6	0	-	1	0.8	1	0.3	1	0.2	2	0.8	3	0.4	-	1.0 :1	2.0 :1
Microphthalmos	8	3.7	5	3.8	13	3.7	14	8.5	7	5.6	21	7.3	22	5.4	12	4.5	34	5.1	0.6 :1	0.7 :1	0.6 :1
All small eyes	9	4.2	6	4.6	13	4.3	14	8.5	8	6.4	23	7.6	23	5.6	14	5.3	37	5.5	0.6 :1	0.7 :1	0.6 :1
Buphthalmos	18	8.4	14	10.5	32	9.2	19	11.6	10	8.0	29	10.0	43	10.6	25	9.5	68	10.2	0.9 :1	1.4 :1	1.1 :1
Glaucoma	0	-	1	0.8	1	0.3	0	-	2	1.6	2	0.7	0	-	3	1.1	3	0.4	-	-	-
Eye removed	0	-	3	2.3	3	0.9	0	-	0	-	0	-	0	-	3	1.1	3	0.4	-	-	-
Disorganised	0	-	2	1.5	2	0.6	0	-	0	-	0	-	0	-	2	0.8	2	0.3	-	-	-
Other	0	-	1	0.8	1	0.3	0	-	0	-	0	-	0	-	1	0.4	1	0.1	-	-	-
Subtotal	28	13.1	35	26.3	63	18.2	34	20.7	31	24.8	65	22.5	69	17.0	67	25.4	136	20.3	0.8 :1	1.1 :1	1.0 :1
Cornea																					
Staphyloma	0	-	1	0.8	1	0.3	0	-	0	-	0	-	0	-	1	0.4	1	0.1	-	-	-
Scarring	4	-	4	3.0	8	2.3	0	-	7	5.6	7	2.4	4	1.0	12	4.5	16	2.4	-	0.6 :1	1.1 :1
Dystrophy	0	-	4	-	4	1.2	0	-	0	-	0	-	0	-	4	1.5	4	0.6	-	-	-

Table 16-1/Conti...	West Bank						Gaza Strip						All Series						WB/GS Ratio		
Anatomical Site	<16		16+		Total WB		<16		16+		Total GS		<16		16+		All ages		<16	16+	All
Other opacities	0	-	1	-	1	0.3	0	-	0	-	0	-	0	-	1	0.4	1	0.1	-	-	-
Subtotal	4	1.9	10	7.5	14	4.0	0	-	7	5.6	7	2.4	4	1.0	18	6.8	22	3.3	-	1.4 :1	2.0 :1
Lens																					
Cataract ^a	48	22.4	14	10.5	62	17.9	34	20.7	24	19.2	58	20.1	94	23.2	38	14.4	132	19.7	1.4 :1	0.6 :1	1.1 :1
Other	9	4.2	0	-	9	2.6	2	1.2	2	1.6	4	1.4	11	2.7	2	0.8	13	1.9	-	-	2.3 :1
Subtotal	57	26.6	14	10.5	90	25.9	36	22.0	26	20.8	69	23.9	105	25.9	40	15.2	145	21.7	1.6 :1	0.5 :1	1.3 :1
Uvea																					
Aniridia	0	-	0	-	0	-	0	-	0	-	0	-	0	-	0	-	0	-	-	-	-
Coloboma	4	1.9	1	0.8	5	1.4	0	-	0	-	0	-	4	1.0	2	0.8	6	0.9	-	-	-
Uveitis	0	-	0	-	0	-	0	-	0	-	0	-	0	-	0	-	0	-	-	-	-
Subtotal	4	1.9	1	0.8	5	1.4	0	-	0	-	0	0.0	4	1.0	2	0.8	6	0.9	-	-	-
Retina																					
Dystrophy	80	37.4	59	44.4	139	40.1	64	39.0	37	29.6	101	34.9	149	36.8	97	36.7	246	36.8	1.3 :1	1.6 :1	1.4 :1
Albinism	6	2.8	2	1.5	8	2.3	13	7.9	6	4.8	19	6.6	19	4.7	8	3.0	27	4.0	0.5 :1	0.3 :1	0.4 :1
ROP	3	1.4	0	-	3	0.9	1	0.6	0	-	1	0.3	4	1.0	0	-	4	0.6	-	-	3.0 :1
Retinoblastoma	1	0.5	0	-	1	0.3	0	-	0	-	0	-	2	0.5	0	-	2	0.3	-	-	-

Table 16-1/Conti...	West Bank						Gaza Strip						All Series						WB/GS Ratio		
Anatomical Site	<16		16+		Total WB		<16		16+		Total GS		<16		16+		All ages		<16	16+	All
Myopia	5	2.3	3	2.3	8	2.3	7	4.3	9	7.2	16	5.5	12	3.0	12	4.5	24	3.6	0.7 :1	0.3 :1	0.5 :1
R25	1	0.5	3	2.3	4	1.2	0	-	4	3.2	4	1.4	2	0.5	7	2.7	9	1.3	-	0.8 :1	1.0 :1
Other	2	0.9	1	0.8	3	0.9	0	-	0	-	0	-	2	0.5	1	0.4	3	0.4	-	-	-
Subtotal	98	45.8	68	51.1	166	47.8	85	51.8	56	44.8	141	48.8	190	46.9	125	47.3	315	47.1	1.2 :1	1.2 :1	1.2 :1
Optic nerve																					
Optic Atrophy	16	7.5	4	3.0	20	5.8	2	1.2	1	0.8	3	1.0	18	4.4	5	1.9	23	3.4	6.1 :1	4.0 :1	6.7 :1
Hypoplasia	0	-	0	-	0	-	0	-	0	-	0	-	0	-	0	-	0	-	-	-	-
Other	0	-	0	-	0	-	0	-	0	-	0	-	0	-	0	-	0	-	-	-	-
Subtotal	16	7.5	4	3.0	20	5.8	2	1.2	1	0.8	3	1.0	18	4.4	5	1.9	23	3.4	-	-	-
Other not listed	2	0.9	0	0	2	0.6	0		0	0	0	0.0	2	0.5	0	-	2	0.3	-	-	-
Globe normal																					
Refractive error	0	-	0	-	0	-	0	-	1	0.8	1	0.3	0	-	1	0.4	1	0.1	-	-	-
Cortical Blindness	0	-	0	-	0	-	2	1.2	0	-	2	0.7	3	0.7	0	-	3	0.4	-	-	-
CIN ^b	1	0.5	0	-	1	0.3	4	2.4	2	1.6	6	2.1	5	1.2	3	1.1	8	1.2	-	-	0.2 :1
Normal vision	0	-	0	-	0	-	0	-	0	-	0		0	-	1	0.4	1	0.1	-	-	-

Undetermined	1	0.5	1	0.8	2	0.6	0	-	1	0.8	1	0.3	1	0.2	2	0.8	3	0.4	-	-	-
Table 16-1/Conti...	West Bank						Gaza Strip						All Series						WB/GS Ratio		
Anatomical Site	<i><16</i>		16+		Total WB		<i><16</i>		16+		Total GS		<i><16</i>		16+		All ages		<i><16</i>	16+	All
Subtotal	2	0.9	1	0.8	3	0.9	6	3.7	4	3.2	10	3.5	9	2.2	7	2.7	16	2.4	-	-	0.3 :1
Undetermined	3	1.4	0	-	3	0.9	1	0.6	0	-	1	0.3	4	1.0	0	-	4	0.6	-	-	-
Total	214	100	133	100	347	100	164	100	125	100	289	100	405	100	264	100	669	100	-	-	1.2 :1

^a Aphakia included under cataract ^b CIN: Congenital Idiopathic nystagmus Percentages in *italic*

Table 16 -1: Anatomical sub-classification: comparison of numbers and region

16.2 Retina

Retina, the largest category, had 315 cases (47.1%) with primary retinal disorder and 18 cases where it was a secondary pathology. The total number of eyes affected in this category, as both primary and secondary, is 636 eyes. This high proportion is witnessed equally in the WB and GS in both the under 16 and 16+ age groups as seen in Table 16-1. The WB/GS ratio however deviates from that of the population ratio between the two regions at 1.2:1 for both age groups demonstrating a higher occurrence of retinal conditions in the GS. (Figures 16-2a, 2b).

The largest subgroup in the retina category is retinal dystrophy (251 patients), making 77.6% of the retinal category, and 36.8% of the total series) (Figure 16-3). The proportion of retinal dystrophies in the GS, especially in the <16 cohort, is higher than that in the corresponding cohort in the WB.

Whilst WB/G population ratio is 1.78:1, that of retinal dystrophies 1.3:1 in the under 16, and in the 16+ the ratio is 1.6:1 which is only marginally lower than the general population ratio. This indicates a higher incidence of retinal dystrophies in the GS.

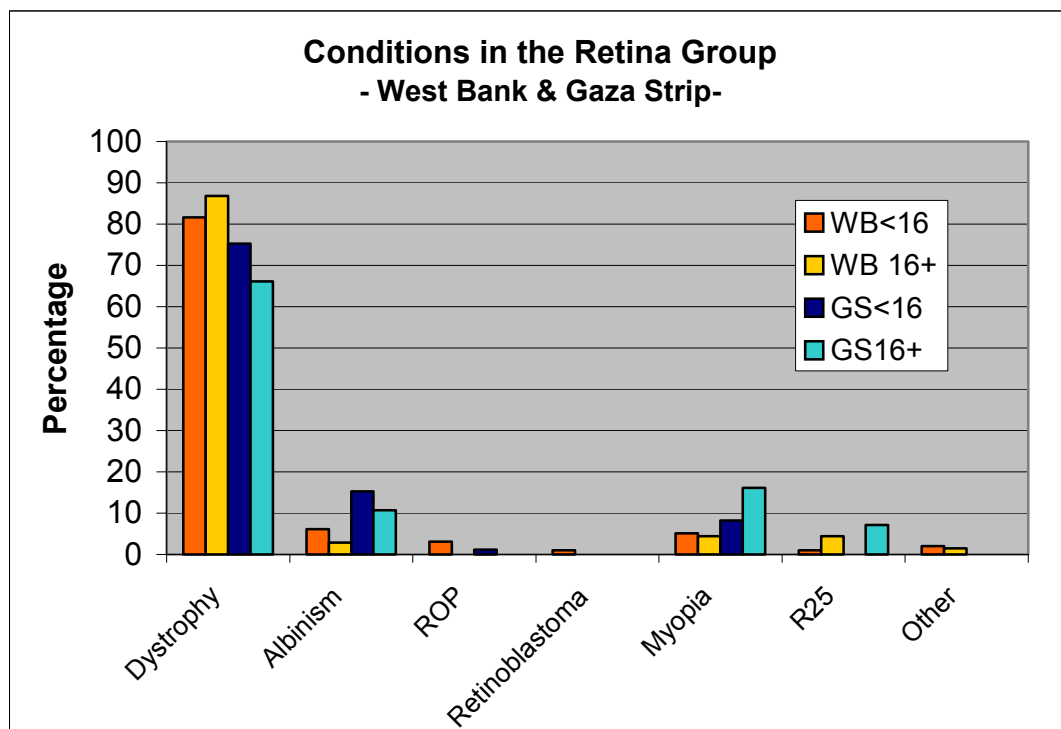


Figure 16 3: Retinal conditions

Retinal dystrophies were followed, in the retinal category, by albinism with 27 patients where albinism was the main pathology, forming 0.6% of the retina category, and 4% of the total series. Albinism forms 6.6% of the total GS cohort but only 2.3% of the total WB conditions

Pathological/syndromatic myopia was the third condition in this category with 25 patients forming 8% of the category and 3.7% of the total series.

Other minor causes were retinopathy of maturity (ROP) with 4 cases forming 0.6% of the series and retinoblastoma with 2 cases forming 0.3% of the series. Both patients with retinoblastoma were absolutely blind, in severe pain from severe proptosis and advanced metastases and died few weeks later (Plates 54, 55).

16.3 Lens (Plates 25-29)

16.3.1 Lens As A Primary Site For Pathology

As an anatomical site for primary pathology, the lens is the second largest category in the total series (n=145) and in the <16 cohort (n=105) forming 21.7% of the total series and 25.9% of the <16. This second place is shared in both the WB

(25.9%) and GS (23.9%). It comes, however, third in the 16+ cohort forming only 15.2% of the series being overtaken by the whole globe (if glaucoma is included in this group).

Gender	WB	GS	Others	All
Males	42	40	8	90
Females	29	22	4	55
Sub-total	71	62	12	145
M/F ratio	1.4:1	1.8:1	2:1	1.6:1

Table 16-2: Gender in lens disorders

There is a preponderance of males in this subgroup and this is more marked in the GS, (Table 16-2). M/F ratios were 1.4:1 and 1.8:1 in the WB and GS respectively with an average ratio of 1.6:1 for the whole series.

16.3.2 Lens Conditions As A Secondary Cause

Lens as the minor anatomical site of pathology was documented in 28 patients with the following categories; whole globe (n=15), retina (n=11) and others (n=2). For clinical conditions, refer to section 16.1.

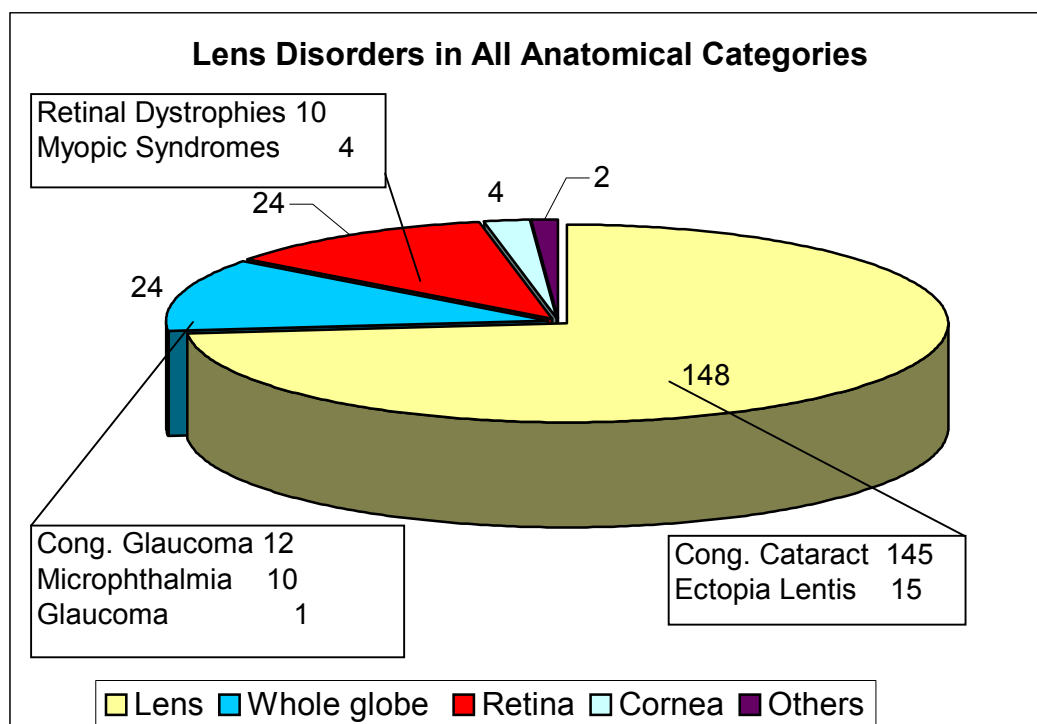


Figure 16-4: Lens disorders in all anatomical categories

16.4 Whole Globe (Plates 21c, 23, 24, 32-35)

This is the third largest anatomical site for pathologies which forms 18.2% and 22.5% of the WB and GS series respectively. A prominent feature of this category is the predominance of the 16+, especially in the WB where they form 26.3% versus 13.1% of the <16. In the GS the figures stand at 24.8% and 20.7% respectively. The former disparity is accounted for by the presence of 7 cases of phthisis, removed and disorganised eyes in the 16+ cohort.

There are three main subcategories in the whole globe anatomical site; these are congenital glaucoma (section 16.4.1), microphthalmia (section 16.4.2), and disorganised eyes (section 16.4.3).

16.4.1 Congenital Glaucoma (Plates 30-31)

The WHO anatomical classification of conditions included buphthalmos in the whole globe category. In more recent literature, this group of conditions has been considered as a separate entity and when classified this way, CG takes from the WG the third position shifting the latter into four-

th place (section 19.2). This category of is the largest in the WG affecting 68 patients and forming 50% of the WG category and 10.2% of the whole series. There were an additional 2 eyes where buphthalmos was a secondary pathology.

16.4.2 Microphthalmia And Anophthalmia (Plates 32-35)

This Who category is the second significant subgroup in the WG affecting 37 patients and this includes 3 cases of anophthalmos, which has been considered in this subgroup. As a primary condition it constitutes 25% of the subcategory and 5.5% of the whole series (Section 19.1).

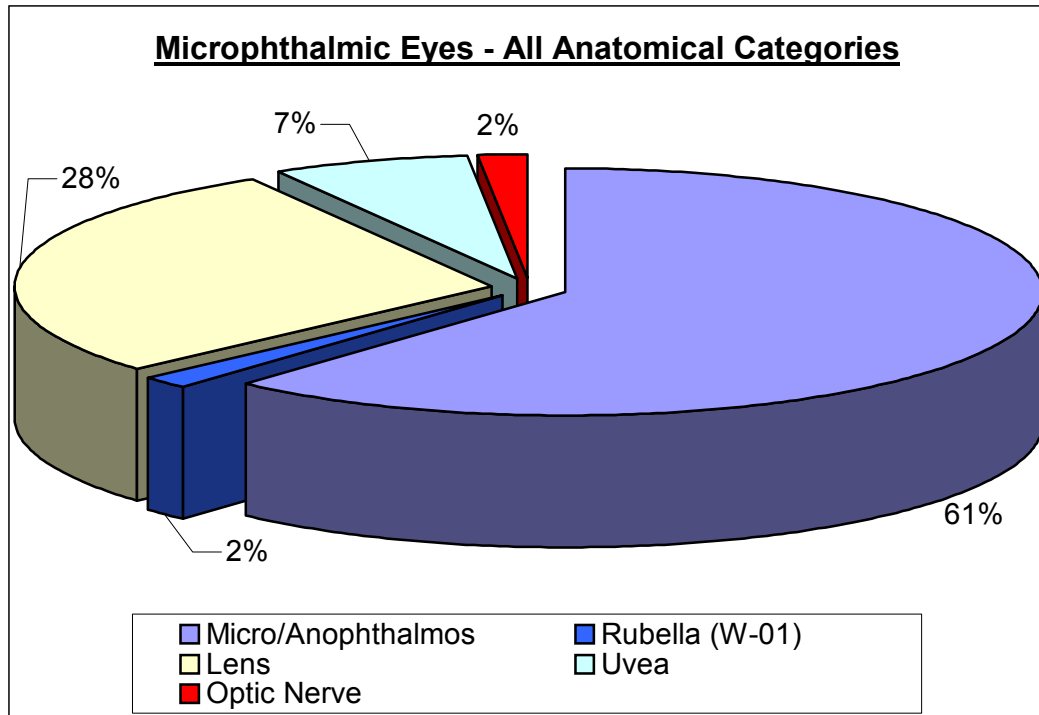


Figure 16-5: Microphthalmia in all anatomical categories

16.4.3 Phthisical, Disorganised And Removed Eyes (Figure 16-6).

Phthisis was present in 18 patients amounting to 2.7% of the total number of eyes (n= 1338). In addition, there were 3 patients with removed eyes and 2 with disorganised eyes making 3.7% of the total series. These had the worse morbidity among the series. Bilateral phthisical eyes were found in 9 patients, 2 of whom had had the other eye enucleated.

Phthisical eyes also existed in another 19 patients who have been classified under different anatomical categories. Figure 16-6 exhibits all the conditions that were associated with phthisical and atrophic eyes both as a primary pathology (in the best seeing eye) and in the other eye. Buphthalmos was the second most important cause of phthisis after the main group which was composed mainly of acquired conditions. Lenticular cases contributed 13% of the total that were congenital cataract and one case of ectopia lentis. The retina's share was 9% with 4 cases, 2 of them were ROP.

As expected, these pathologies show wide variation in incidence with age; it is more prevalent among the 16+ at a ratio of 8.5:1.

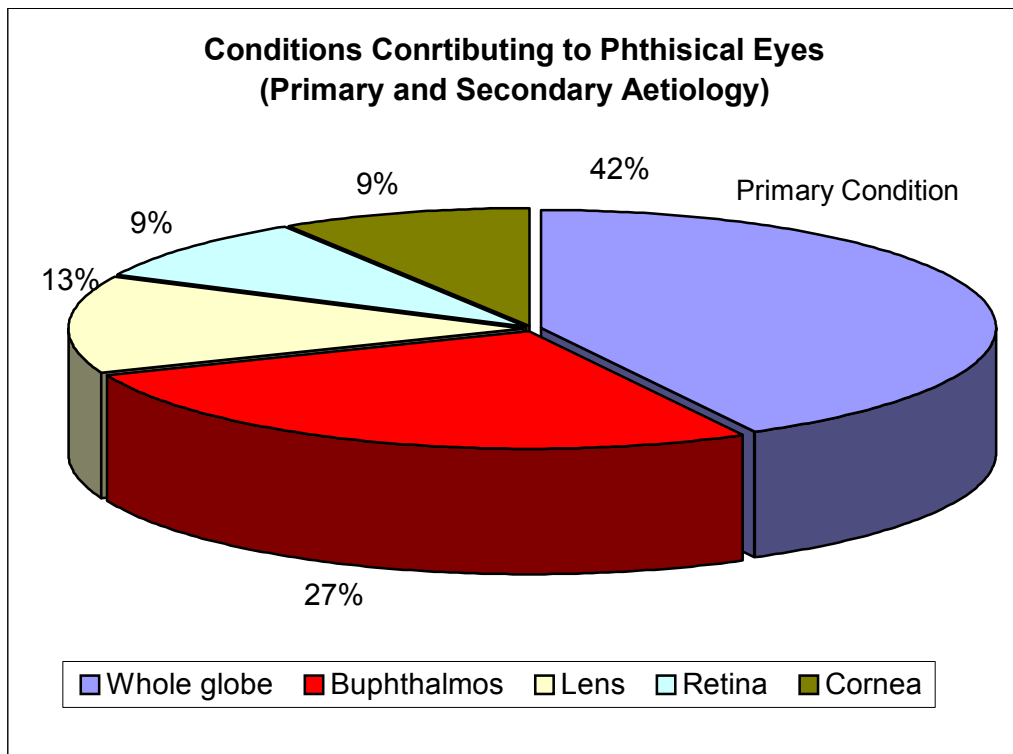


Figure 16-6: Conditions contributing to phthisical eyes

As with the majority of conditions, there is a higher prevalence of phthisis in the GS as seen in the WB/GS ratio of 1:1 for the <16 and 0.8:1 for the 16+ in comparison to the general population ratio of 1.78:1. Out of the 18 cases, 16 were above the age of 16 with an average age of 37 years (range 5-86 years). All the primary phthisical cases were acquired. Almost

three quarters were secondary to infection and post measles and 2 resulted from explosions (Table 16-3).

Distribution of Phthisical/Atrophic Eyes in the West Bank and Gaza Strip		
	<u><16</u>	<u>16+</u>
West Bank	1 (0.5%)	8 (5.3%)
Gaza Strip	1 (0.6%)	9 (7.2%)

Table 16-3:
Phthisical and atrophic eyes by region

16.5 Cornea (Plates 21, 24, 26)

The cornea was the primary anatomical site of pathology in 22 cases (3.3% of the total) of whom only 4 were under the age of 16 years and from the WB, the remaining 18 being adults. The distribution of patients in the two regions and according to age is shown in figure 16-7 below. No cases with bilateral blindness were registered from the GS in the under 16 with corneal conditions.

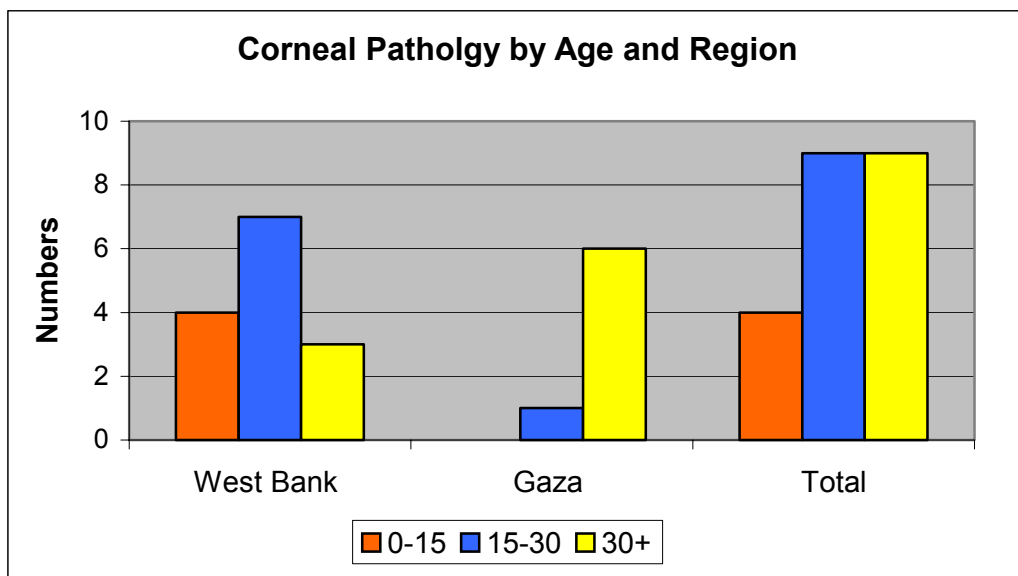


Figure 16-7: Comparison of corneal pathology between the WB and GS

Simultaneous involvement of both eyes by the same corneal pathology was present in 13 cases. In 39 cases the cornea was the secondary anatomical site of pathology.

The main primary pathology of corneal diseases was corneal scarring and opacification which affected 18 patients (2.4% of the total series). Figure 16-8 below highlights the predominance of the corneal conditions in the older age group and its absence among the children in the GS.

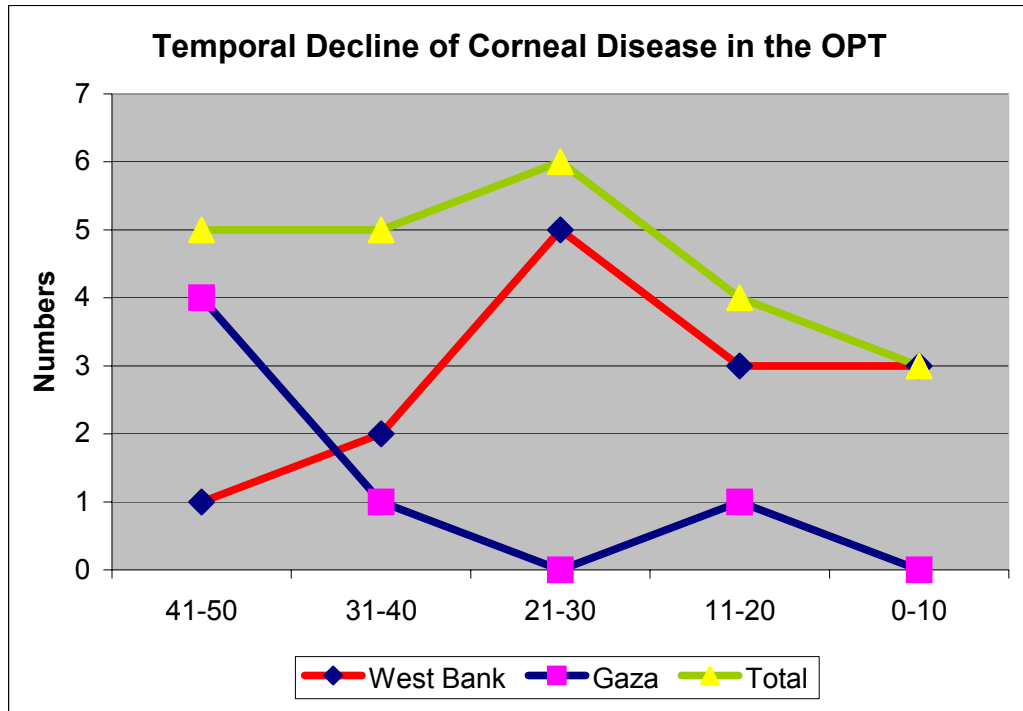


Figure 16-8: Temporal decline of corneal blindness in the OPT

The combined primary and secondary corneal pathologies total 60 patients, 31 of them with corneal scarring. This means that 9% of the whole series had corneal pathology of some kind and nearly half of these were corneal scarring.

16.6 Optic Nerve (Plates 58a, b)

Optic nerve conditions formed a very small category with 23 patients all affected with optic atrophy. There was no optic nerve hypoplasia as a cause of visual loss. The distribution of these cases is delineated in Table 16-9 which shows a significant disparity between the WB and GS with a WB/GS ratio 8:1 in the under 16 cohort. Nearly half of cases are childhood onset (n=11), 6 are hereditary, 3 from prenatal causes, 1 perinatal and 2 are from undetermined causes. The pathologies of the optic nerve disorders are

explained in depth under the non-hereditary causes (section 18 and Table 18-5). Of the 23 cases, 18 are children and 5 adults (Table 16-4).

These conditions come high in their visual morbidity with 87% (n=20/23) in the svi/bl rank (category '2', 3 patients; category '4', 9 patients; category '5', 9 patients; and category '7', 2 patients).

Optic Nerve Conditions			
	<u>WB</u>	<u>GS</u>	<u>B/GS</u>
<16	16	2	8:1
16+	4	1	4:1
Total	20	3	6.6:1

Table 16.4: Distribution of Optic Nerve Conditions

16.7 Uvea (Plates 33, 35)

Uvea was the smallest category of anatomical site for primary pathology with only 6 patients as a primary condition. Uveal and optic nerve head coloboma was the only pathology in this category and formed 0.9% of the total series. It is interesting that all these cases are from the WB and one from an unknown location. No case was reported from GS. (Table 16-5). Visual morbidity in these patients is also poor with two thirds of the patients out of 6 in the svi/bl category (66%, n=4/6).

This is by no means the total number of uveal colobomas in the series as most of these are included in the other anatomical sites such as the whole globe (microphthalmos) and lens (Congenital cataract) totalling 19 patients. Of these 19 cases, 17 are from WB and GS.

In the former, 7 were under 16 and in the latter, only one. The distribution is shown in the Table 16-5 and figure 16-9. Clinically, uveal coloboma existed in different combinations

ranging from total (iris, choroidal and ONH in 3, iris involvement in 8, and optic disc involvement in 11.

	<u>WB</u>	<u>GS</u>	<u>Series</u>
<16	5	0	5
16+	0	0	1
Total	5	0	6

Table 16-5: Uveal pathology by region

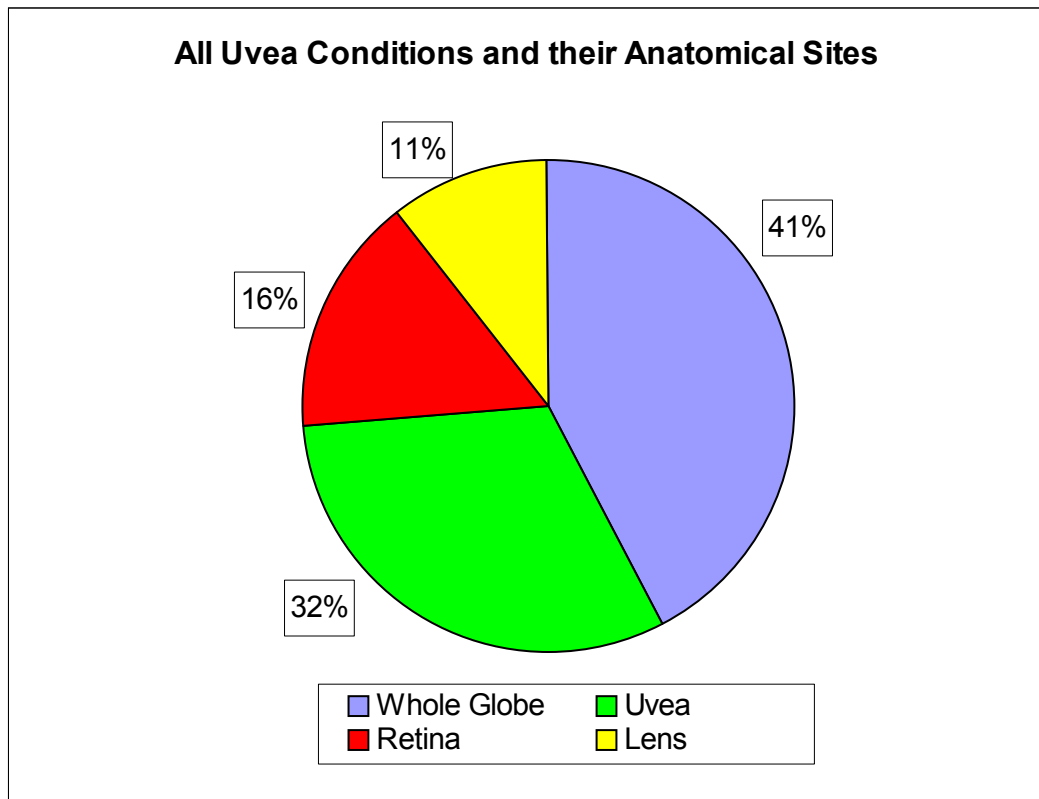


Figure 16-9: Anatomical sites of all uveal pathology

16.8 ‘Globe Appears Normal’ Category (Table 16-6)

This is a small category with 16 patients. The main group of conditions is congenital idiopathic nystagmus (n=8). Cortical blindness contributed to 3 patients, refractive error to 1, and the rest were mainly unidentified conditions. Idiopathic nystagmus is predominantly a GS condition in view of the presence of an extended pedigree with several sibships with this condition.

The second condition in this category is cortical blindness with 2 patients from GS and one unidentified. There are another 5 cases in this category including undetermined and refractive error.

16.9 Other Categories

The other two categories of the anatomical classification are the ‘Others Not Listed’ and the ‘Undetermined’ with 2 and 4 patients respectively.

	Normal Appearing Globe		
	<16	16+	All Ages
Idiopathic Nystagmus			
West Bank	1	0	1
Gaza Strip	2	4	6
Others	0	1	1
Total	5	3	8
Cortical Blindness			
West Bank	2	-	2
Gaza Strip	1	-	1
Total	3	-	3

Table 16-6: Distribution of 'Normal Appearing Globe' WHO category