

# Anatomical Classifications

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## Introduction

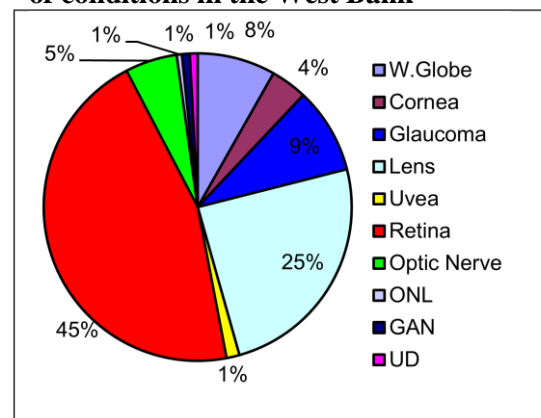
Based on the protocol outlined in the WHO/PBL Eye Examination Record for Children with Blindness and Low Vision, Coding Instruction, conditions were reclassified anatomically and by underlying cause. Where there was a difference in the anatomical diagnoses between the two eyes, then the diagnosis of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

### 14.1 Anatomical Sites

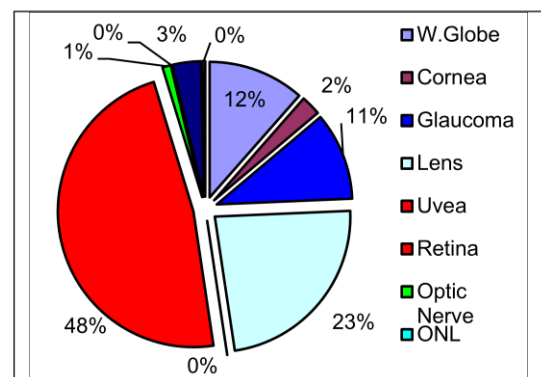
In this survey, the main anatomical sites of pathology are the retina, the lens, and the whole globe. When glaucoma is analysed separately rather than part of the whole globe, it takes the third place followed by the rest of the whole globe. (Figures 14.1, 14.2)

Details of anatomical sites of the primary pathologies are found in Chapter 14 Appendix (Table A14.1 and Figure A14.1)

**Figure 14.1 Anatomical classification of conditions in the West Bank**



**Figure 14.2 Anatomical classification of conditions in the Gaza Strip**



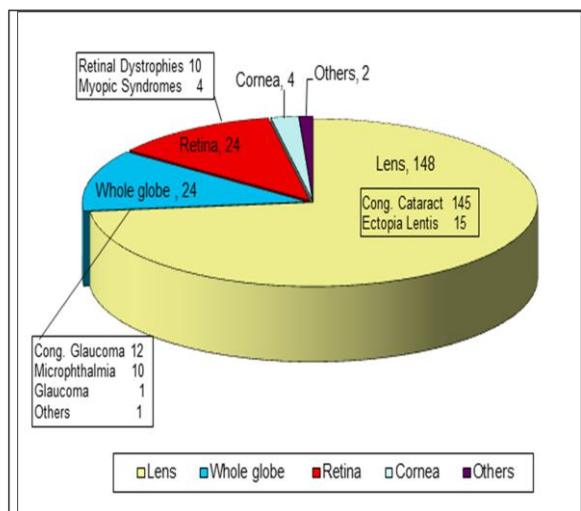
## 4.2 Lens

### 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). (Figure 14.3)

There is a preponderance of males in this subgroup and this is more marked in the GS. (Figure 14.1; Table 14.1). 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.

**Figure 14.3 Lens disorders in all anatomic categories**



### Lens Conditions as a Secondary Cause

Lens as a 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).

**Table 14.1 Gender in lens disorders**

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

## 14.3 Whole Globe

This is the third largest anatomical site for pathologies and 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, microphthalmia, and disorganised eyes.

## 14.4 Congenital Glaucoma

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

### 14.5 Small Eyes

This WHO category is the second significant subgroup in the WG affecting 37 patients and this includes 3 cases of anophthalmos, which have been considered in this subgroup. As a primary condition it constitutes 25% of the subcategory and 5.5% of the whole series. Table 14.3 shows comparable regional figures.

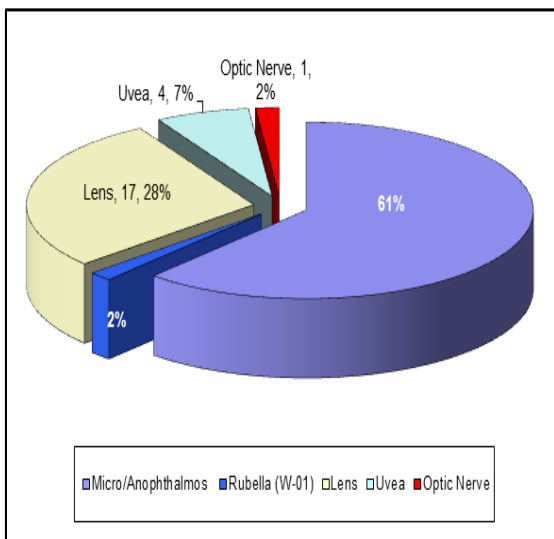
#### Phthisical, Disorganised / Removed Eyes

Phthisis was present in 18 patients amounting to 2.7% of the total number of eyes (n= 1338). In addition, 3 patients had their eyes removed and in 2 the eye was disorganised making 3.7% of the total series. These had the worst morbidity in the series. Bilateral phthisical eyes were found in 9 patients, 2 of whom had had the other eye enucleated. (Figure 14.5)

**Table 14.2 Phthisical /atrophic eyes by region**

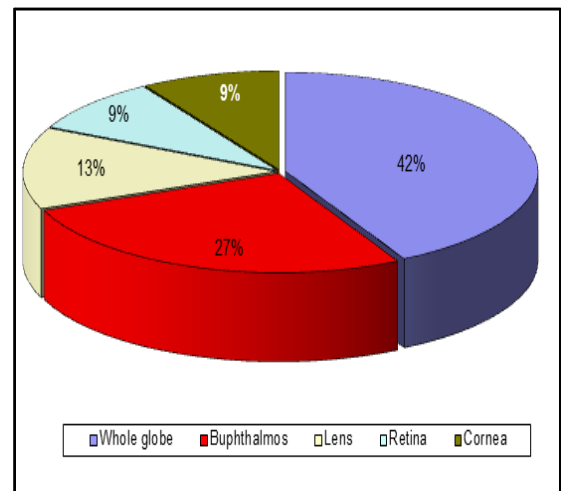
	<u>&lt;16 years</u>	<u>16+</u>
<b>West Bank</b>	<b>1 (0.5%)</b>	<b>8 (5.3%)</b>
<b>Gaza Strip</b>	<b>1 (0.6%)</b>	<b>9 (7.2%)</b>

**Figure 14.4 Small eyes in all anatomical categories**



Phthisical eyes also existed in another 19 cases who have been classified under different anatomical categories. Figure 14.5 demonstrates 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 phthisical eyes which were essentially congenital cataract and one case of ectopia lentis which became phthisical following trauma. The retina share was 9% with 4 cases, 2 of which were ROP. (Figure 14.5)

**Figure 14.5 Conditions contributing to phthisical eyes (as primary and secondary conditions causing blindness)**



These pathologies show wide variations in incidence with age; it is more prevalent among the 16+ at a ratio of 8.5:1.

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 two resulted from explosions (Chapter 15).

### 14.6 Cornea

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 14.6 below. No cases with bilateral blindness were registered from the GS in the <16 with corneal conditions. 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

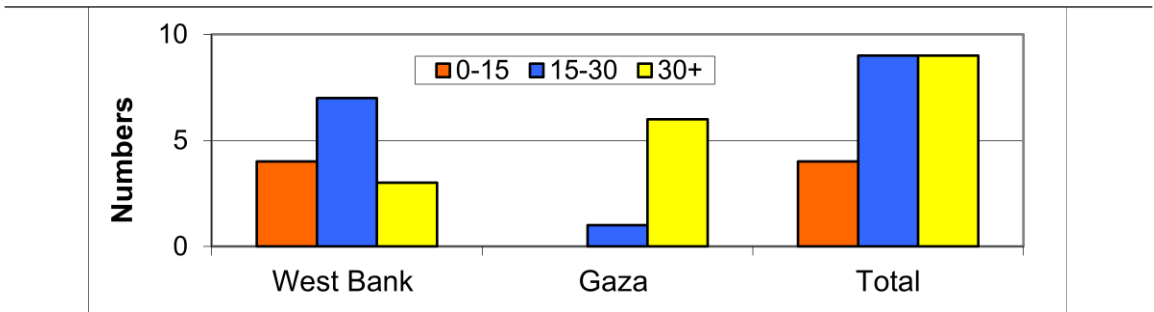
diseases was corneal scarring/ opacification which affected 18 patients (2.4% of the total series). Figure 14.6 below highlights the predominance of the corneal conditions in the older age group and its absence among children in the GS. (Figure 14.7)

The combined primary and secondary corneal pathologies totalled 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.

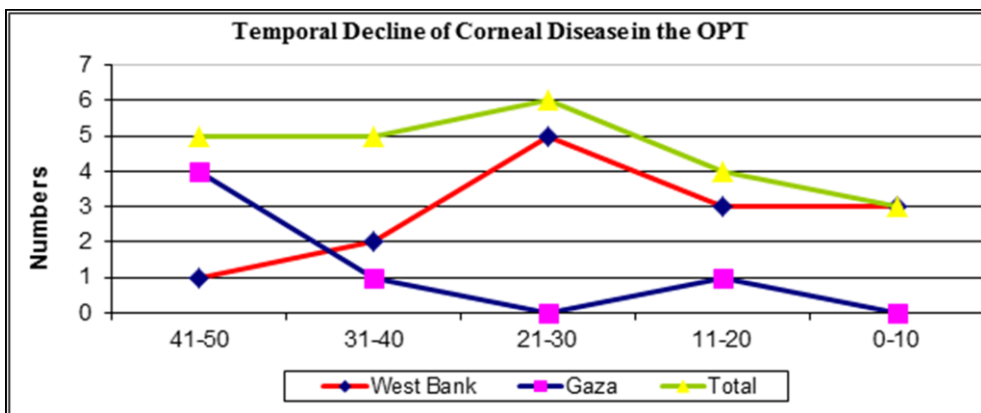
### 14.7 Optic Nerve

Optic nerve conditions formed a very small category with only 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

**Figure 14.6 Comparison of corneal pathology between the West Bank and Gaza Strip by age and region**



**Figure 14.7 Temporal decline in acquired corneal pathologies**



14.9 which shows a significant disparity between the WB and GS with a WB/GS ratio 8:1 in the under 16 cohort. 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 14.9 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 14.3)

**Table 14.3 Distribution of Optic Nerve Conditions by age and region**

	West Bank	Gaza Strip	Both
<16	16	2	18
16+	4	1	5
Total	20	3	23

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

## 14.8 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. (Figure 14.3) 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 14.1, 14.2)

The largest subgroup in the retinal category is retinal dystrophy (251 patients), comprising

77.6% of the retinal category, and 36.8% of the total series) (Figure 14.3). 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/GS 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.

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

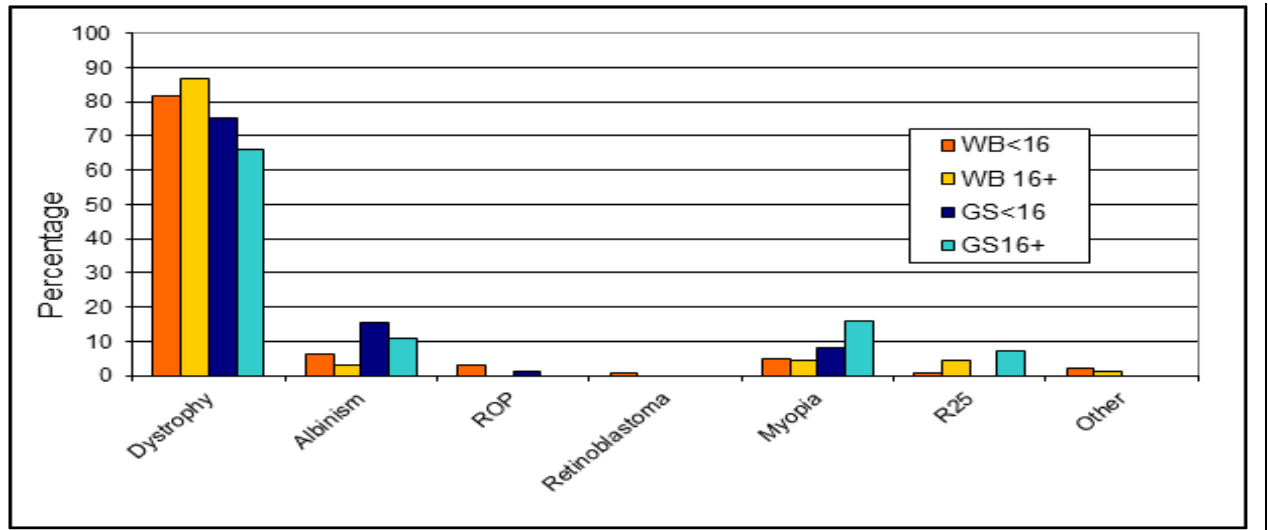
Pathological/syndromatic myopia was the third condition in this category with 25 patients forming 8% of the category and 3.7% of the total series.

Other minor causes were retinopathy of prematurity (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.

## 14.9 Uvea

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 14.4). 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).

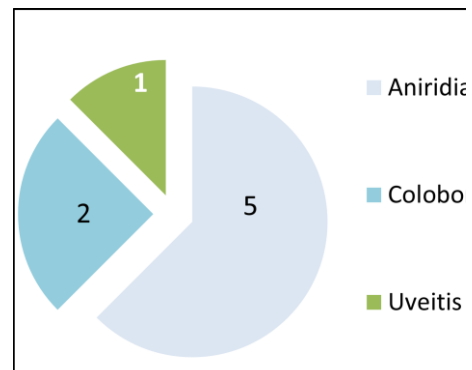
**Figure 14.3** Retinal the West Bank and Gaza Strip



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 < 16 and in the latter, only one. (Table 14.5; Figure 14.9) Clinically, uveal coloboma existed in different combinations ranging from total iris, choroidal and optic nerve hypoplasia in 3, iris involvement in 8, and optic disc involvement in 11.

**Table 14.5** Uveal pathologies



**Table 14.4** Uveal pathology by age and region

	West Bank	Gaza Strip	Both
<16	5	0	5
16+	0	0	1
Total	5	0	6

**14.10 Normal Looking Globe Category**

This is a small category with 16 patients. (Table 14.6). The main group of conditions is congenital idiopathic nystagmus (n=8). Cortical blindness contributed to 3 patients, refractive error to 1, and the remainder 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.

**Table 14.6 Distribution of ‘Normal Appearing Globe’ WHO category**

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

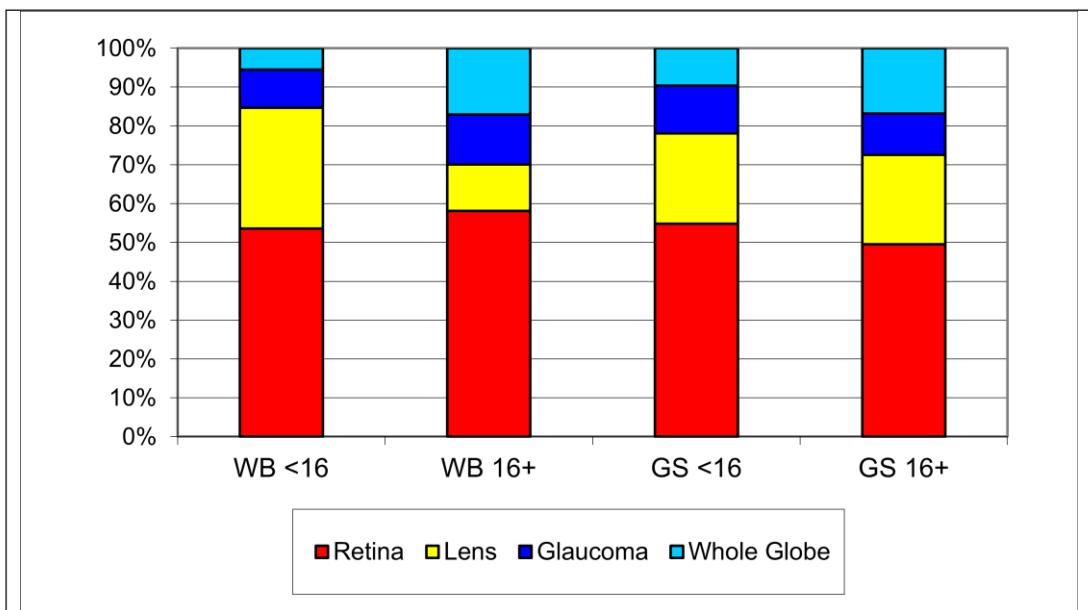
The second condition in this category is cortical blindness with 2 patients from GS and one unidentified. There are another 5 cases in this category including undetermined and refractive error.

**14.11 Other Categories**

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

**APPENDIX CHAPTER 14**

**Figure A14.1 Main anatomical sites of pathology by region and age cohorts**



**Table A14.2 Anatomical classification of pathologies; numbers by region and age cohort**

Anatomical Site	West Bank			Gaza Strip			Both Regions			WB:GS Ratio					
	<16	16+	Total	<16	16+	Total	<16	16+	Total	<16	16+	Total			
<b>Whole Globe</b>															
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
<b>Cornea</b>															
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	-	-	-
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



<b>Lens</b>																								
Cataract <sup>a</sup>	48	<b>22.4</b>	14	<b>10.5</b>	62	<b>17.9</b>	34	<b>20.7</b>	24	<b>19.2</b>	58	<b>20.1</b>	94	<b>23.2</b>	38	<b>14.4</b>	132	<b>19.7</b>	1.4	:1	0.6	:1	1.1	:1
Other	9	<b>4.2</b>	0	-	9	<b>2.6</b>	2	<b>1.2</b>	2	<b>1.6</b>	4	<b>1.4</b>	11	<b>2.7</b>	2	<b>0.8</b>	13	<b>1.9</b>	-	-	-	-	2.3	:1
Subtotal	57	<b>26.6</b>	14	<b>10.5</b>	90	<b>25.9</b>	36	<b>22.0</b>	26	<b>20.8</b>	69	<b>23.9</b>	105	<b>25.9</b>	40	<b>15.2</b>	145	<b>21.7</b>	1.6	:1	0.5	:1	1.3	:1
<b>Uvea</b>																								
Aniridia	0	-	0	-	0	-	0	-	0	-	0	-	0	-	0	-	0	-	-	-	-	-	-	-
Coloboma	4	<b>1.9</b>	1	<b>0.8</b>	5	<b>1.4</b>	0	-	0	-	0	-	4	<b>1.0</b>	2	<b>0.8</b>	6	<b>0.9</b>	-	-	-	-	-	-
Uveitis	0	-	0	-	0	-	0	-	0	-	0	-	0	-	0	-	0	-	-	-	-	-	-	-
Subtotal	4	<b>1.9</b>	1	<b>0.8</b>	5	<b>1.4</b>	0	-	0	-	0	<b>0.0</b>	4	<b>1.0</b>	2	<b>0.8</b>	6	<b>0.9</b>	-	-	-	-	-	-
<b>Retina</b>																								
Dystrophy	80	<b>37.4</b>	59	<b>44.4</b>	139	<b>40.1</b>	64	<b>39.0</b>	37	<b>29.6</b>	101	<b>34.9</b>	149	<b>36.8</b>	97	<b>36.7</b>	246	<b>36.8</b>	1.3	:1	1.6	:1	1.4	:1
Albinism	6	<b>2.8</b>	2	<b>1.5</b>	8	<b>2.3</b>	13	<b>7.9</b>	6	<b>4.8</b>	19	<b>6.6</b>	19	<b>4.7</b>	8	<b>3.0</b>	27	<b>4.0</b>	0.5	:1	0.3	:1	0.4	:1
ROP	3	<b>1.4</b>	0	-	3	<b>0.9</b>	1	<b>0.6</b>	0	-	1	<b>0.3</b>	4	<b>1.0</b>	0	-	4	<b>0.6</b>	-	-	-	-	3.0	:1
Retinoblastoma	1	<b>0.5</b>	0	-	1	<b>0.3</b>	0	-	0	-	0	-	2	<b>0.5</b>	0	-	2	<b>0.3</b>	-	-	-	-	-	-
Myopia	5	<b>2.3</b>	3	<b>2.3</b>	8	<b>2.3</b>	7	<b>4.3</b>	9	<b>7.2</b>	16	<b>5.5</b>	12	<b>3.0</b>	12	<b>4.5</b>	24	<b>3.6</b>	0.7	:1	0.3	:1	0.5	:1
R25	1	<b>0.5</b>	3	<b>2.3</b>	4	<b>1.2</b>	0	-	4	<b>3.2</b>	4	<b>1.4</b>	2	<b>0.5</b>	7	<b>2.7</b>	9	<b>1.3</b>	-	-	0.8	:1	1.0	:1
Other	2	<b>0.9</b>	1	<b>0.8</b>	3	<b>0.9</b>	0	-	0	-	0	-	2	<b>0.5</b>	1	<b>0.4</b>	3	<b>0.4</b>	-	-	-	-	-	-
<b>Optic nerve</b>																								
Optic Atrophy	16	<b>7.5</b>	4	<b>3.0</b>	20	<b>5.8</b>	2	<b>1.2</b>	1	<b>0.8</b>	3	<b>1.0</b>	18	<b>4.4</b>	5	<b>1.9</b>	23	<b>3.4</b>	6.1	:1	4.0	:1	6.7	:1

Hypoplasia	0	-	0	-	0	-	0	-	0	-	0	-	0	-	-	-	-				
Other	0	-	0	-	0	-	0	-	0	-	0	-	0	-	-	-	-				
Subtotal	16	<b>7.5</b>	4	<b>3.0</b>	20	<b>5.8</b>	2	<b>1.2</b>	1	<b>0.8</b>	3	<b>1.0</b>	18	<b>4.4</b>	5	<b>1.9</b>	23	<b>3.4</b>	-	-	-
<b>Other not listed</b>																					
	2	<b>0.9</b>	0	<b>0</b>	2	<b>0.6</b>	0		0	<b>0</b>	0	<b>0.0</b>	2	<b>0.5</b>	0	-	2	<b>0.3</b>	-	-	-
<b>Normal Looking Globe</b>																					
Refractive error	0	-	0	-	0	-	0	-	1	<b>0.8</b>	1	<b>0.3</b>	0	-	1	<b>0.4</b>	1	<b>0.1</b>	-	-	-
Cortical Blindness	0	-	0	-	0	-	2	<b>1.2</b>	0	-	2	<b>0.7</b>	3	<b>0.7</b>	0	-	3	<b>0.4</b>	-	-	-
CIN <sup>b</sup>	1	<b>0.5</b>	0	-	1	<b>0.3</b>	4	<b>2.4</b>	2	<b>1.6</b>	6	<b>2.1</b>	5	<b>1.2</b>	3	<b>1.1</b>	8	<b>1.2</b>	-	-	0.2 :1
Normal vision	0	-	0	-	0	-	0	-	0	-	0		0	-	1	<b>0.4</b>	1	<b>0.1</b>	-	-	-
Undetermined	1	<b>0.5</b>	1	<b>0.8</b>	2	<b>0.6</b>	0	-	1	<b>0.8</b>	1	<b>0.3</b>	1	<b>0.2</b>	2	<b>0.8</b>	3	<b>0.4</b>	-	-	-
Subtotal	2	<b>0.9</b>	1	<b>0.8</b>	3	<b>0.9</b>	6	<b>3.7</b>	4	<b>3.2</b>	10	<b>3.5</b>	9	<b>2.2</b>	7	<b>2.7</b>	16	<b>2.4</b>	-	-	0.3 :1
<b>Undetermined</b>																					
	3	<b>1.4</b>	0	-	3	<b>0.9</b>	1	<b>0.6</b>	0	-	1	<b>0.3</b>	4	<b>1.0</b>	0	-	4	<b>0.6</b>	-	-	-
<b>Total</b>	214	<b>100</b>	133	<b>100</b>	347	<b>100</b>	164	<b>100</b>	125	<b>100</b>	289	<b>100</b>	405	<b>100</b>	264	<b>100</b>	669	<b>100</b>	-	-	1.2 :1

Percentages in **bold italic**. <sup>a</sup> Aphakia included under cataract <sup>b</sup> CIN: Congenital Idiopathic nystagmus

