

18 NON-HEREDITARY CONDITIONS

18.1 Non-Hereditary Causes

18.1.1 Distribution

Number of patients with non-genetic conditions in the total series of 669 patients is 90, forming 13.3% of the series. Fifty five of those come from WB forming 15.9% of the series and 29 from the GS, 10% of the series. This distribution constitutes a ratio of 1.9:1, slightly higher than the population ratio between the two regions of 1.78:1. Table 17-1, 2 and 3 illustrate the distribution of the pedigrees, sibships, and patients among the hereditary and non-hereditary disorders.

There is a preponderance of adults in this category with a <16/16+ ratio of 0.8:1 in contrast to the same ratio in the hereditary condition which is 1.48:1. The sex ratio in this group (1.19:1) is comparable to the ratio in hereditary conditions.

18.2 Factors in Non-Hereditary Conditions

The causation of the 90 cases of non-hereditary conditions span the whole spectrum of non-hereditary factors as follows (Figure 18-1): -

- A) Prenatal factors with 24 patients (27%).
- B) Perinatal factors (including neonatal) with 10 patients (11%).
- C) Postnatal factors, the largest group with 55 patients (61.8%).
- D) Uncertain non-genetic, 1 patient.

18.3 Pathologies in the Non-Hereditary Conditions

Whole globe pathologies formed the main group of conditions in the non-hereditary conditions with 31 patients constituting 35% of cases. This is followed by corneal conditions comprising 16 cases (18%) and thirdly by

optic nerve disorders with 14 cases (16%), whilst lens pathologies had 11 cases (12%) and retinal conditions 9 cases (10%).

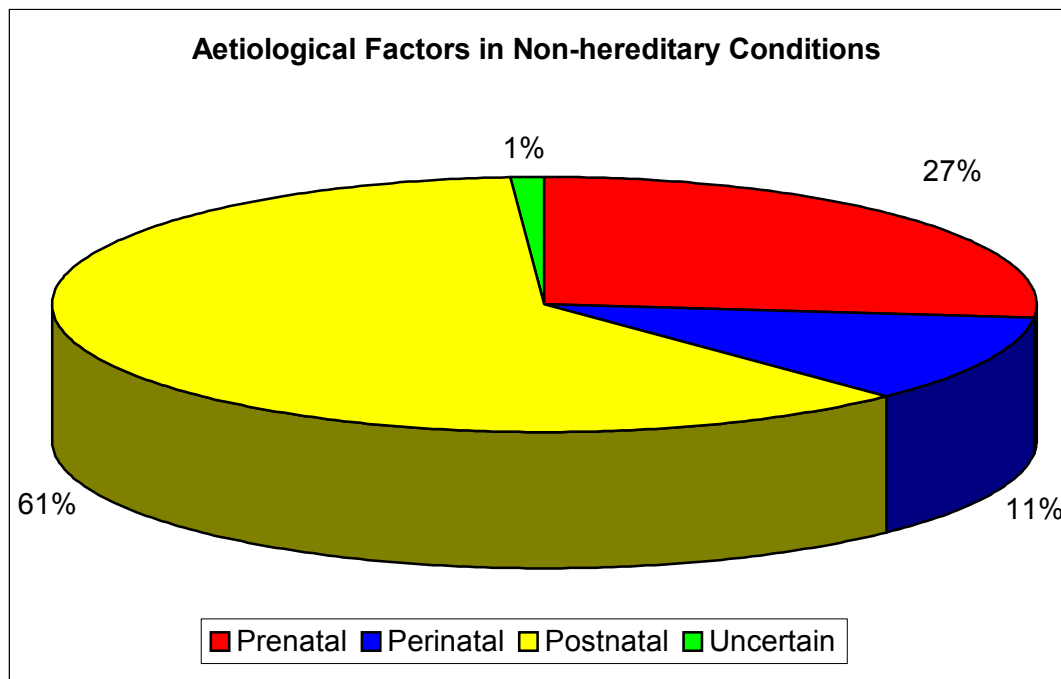


Figure 18-1: Aetiological factors in non-hereditary conditions

Table 18-1 gives the number of cases and percentages of each of the non-hereditary subgroup.

18.4 Prenatal Factors

18.4.1 Causations

The contributory factors in the patients studied are enumerated in the box below.

Among the 24 cases of prenatal aetiologies, the majority are most probably secondary to intrauterine infection constituting 79% of the prenatal cases and making this factor the most important cause for prenatal conditions which amounts to 3.6% of the total series of 669. In most cases of the undetermined type of intra uterine infections, it was not possible to ascertain the exact cause although CMV virus infection is a likely possibility. There were 3 cases with possible drug intake in the first trimester (Table 18-2).

	No.	% Subgroup	% Total
Prenatal (28%)			
Lens	11	45.8	1.6
Whole globe	8	33.3	1.1
Optic nerve	3	12.5	0.4
Others	3	8.3	0.3
Subtotal	25	100	3.4
Perinatal (11%)			
Retina	4	40.0	0.6
Cornea	2	20.0	0.3
Phthisis	2	20.0	0.3
Others	2	20.0	0.3
Subtotal	10	100	1.4
Postnatal (61%)			
Whole globe ^a	21	38.2	3.0
Cornea	14	25.5	2.0
Optic atrophy	11	20.0	1.6
Retinal causes	5	9.1	0.7
Others	4	7.3	0.6
Subtotal	55	100	7.9
Uncertain (1.1%)			
	1		0.1
Total	90		12.9

^a Phthisis 10 cases

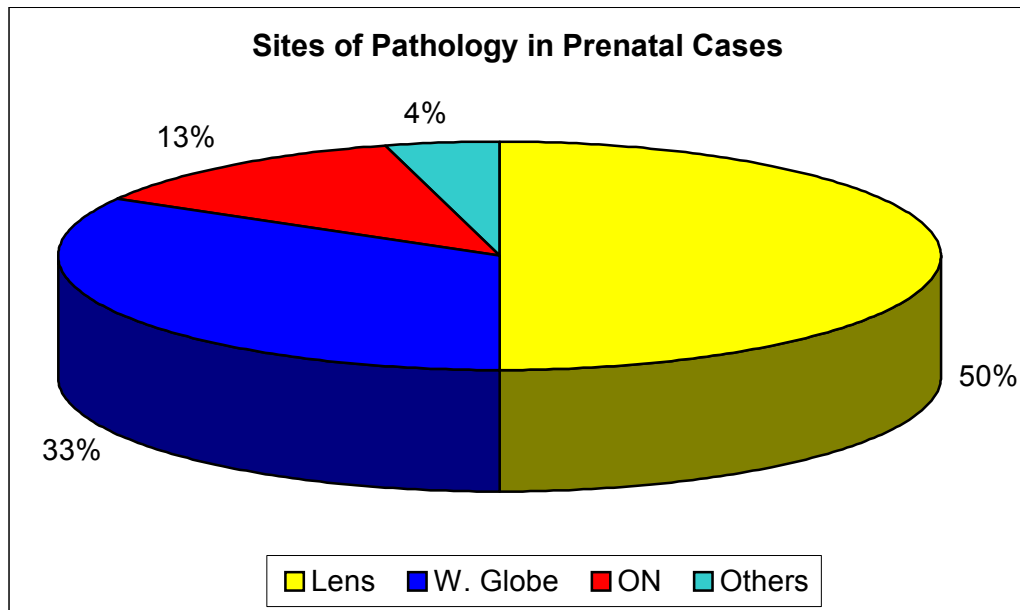
Table 18-1: Non-hereditary factors by anatomical site of

<u>Intrauterine Causes</u>	
1. Intrauterine infections (IUI)	
a. Congenital Toxoplasmosis	3
b. Congenital Rubella	1
c. Undetermined type of IUI	10
2. Possible IUI	5
3. Possible drug intake	3
4. Either IUI or drug intake	1
5. Undetermined IU Factors	<u>1</u>
	Total 24

Table 18-2: Intrauterine causes

18.3.2 Gender in Prenatal Conditions

See section 14.3.3.



18.3.3 Anatomical Sites

The lens was the main site of pathology was found in 50% of the prenatal cases, followed by the whole globe in 33%. The main clinical conditions in this subgroup are CC followed by microphthalmos.

Visual acuities in the prenatal group of patients were severely compromised with 71% (n= 17) blind with nearly two thirds of the blind having NLP. The visual acuities in cases with lenticular aetiology varied between categories '2' and '3' (Table 18-3).

There is a significant preponderance of prenatal conditions in the WB versus GS with a ratio of 15:1.

Category '5' (NLP)	10
Category '4' (blind)	7
Category '3' (svi)	1
Category '2' (vi)	3
Category '1' (no vi)	1
Others	2
Total	24

Table 18-3: Visual acuities in prenatal cases

18.4 Perinatal and Neonatal Causes

This is a very small group consisting of 10 patients only. Four of these patients were in the WB and three in the GS. The number of females in the GS was double that of males (2 and 1). Two thirds (n=4) of the perinatal problems were caused by ROP and the other third (n=2) were from birth hypoxia (Figure 18-3).

There were only 4 cases where the cause of blindness was due to factors occurring in the first month after birth. One was described as due to ophthalmia, two due to harmful topical medications (one of which was iatrogenic as a result of a medical error), and the fourth case was secondary to surgical complications in the first month (neither the nature of the condition nor of the surgery were clear).

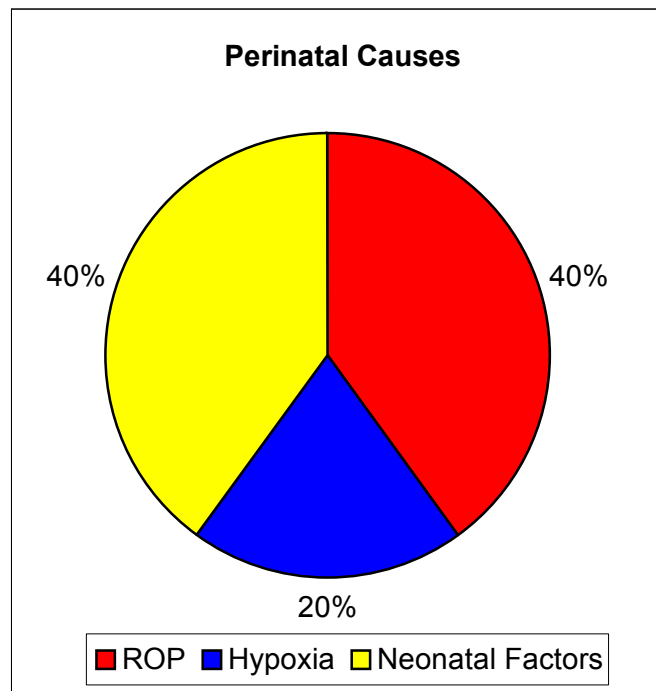


Figure 18-3: Perinatal causes

Anatomical site of the pathologies was mainly the retina in 4 (n=6). Two cases were phthisical, 2 had corneal pathology and 1 optic OA.

Visual acuities were also severely affected in this category with 100% blind/svi (5 with NLP, 3 blind category 4, 1 with svi, and 1 in category '7') (Figure 18-4).

18.5 Postnatal Factors

(Plates 21-24)

This is the largest group of the acquired non-genetic cases with 55 patients, forming 62% of the non-hereditary conditions and 8% of the total series. Twenty of these patients were under the age of 16 (36%) and 35 (64%) are adults. This group comprised 31 patients from the WB as opposed to 21 in the GS, thus producing a ratio of 1.5:1, which was fairly similar to the population ratio, taking into account the small size of the sample and the possible presence of a few scattered cases in the WB that had not been examined (population ratio 1.78:1). There were an additional 2 where the origin was not determined and one from Israel.

The M/F ratio in the various age cohorts in this group shows that with the exception of 16+ cohort in the WB, there is a male preponderance the highest being in the GS <16 cohort at 4:1, followed by the WB <16 cohort at 2.3:1 and finally the GS <16 cohort at 1.6:1. The WB 16+ cohort showed female preponderance with a ratio of 0.36:1.

In this aetiological group, 40% of cases belonged to the whole globe (n=22), 25.5% to the cornea (n=14), 20% to the optic nerve (n=11) and 9% to the retina (n=5). The 14 corneal cases were corneal scarring and 1 was staphyloma. Eleven of these were 16+ and 3 <16. In the whole globe cases, 12 were phthisical/atrophic eyes and in 3, eyes had been removed.

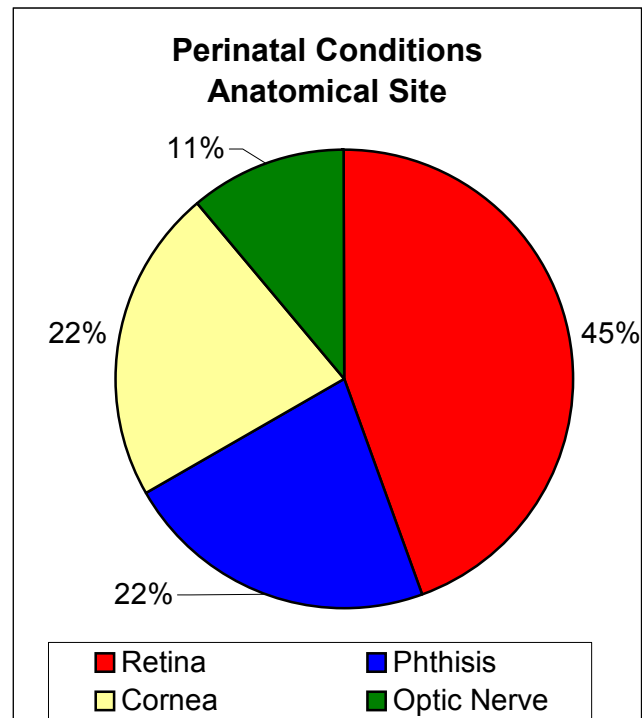


Figure 18-4: Anatomical sites of pathology in perinatal conditions

Conditions	Nos.	% Sub- group	% Total
Prenatal (Intrauterine) Causes			
<u>Intrauterine infection (IUI)</u>			
Congenital toxoplasmosis	3	12	3.3
Congenital rubella	1	4	1.1
Undetermined	10	40	11
Intrauterine infection suspect	5	20	5.5
Intrauterine medicine suspect	3	12	3.3
<u>Undetermined IU factors</u>	3	12	3.3
Subtotal IUI	25	100	27.7
Perinatal Causes			
ROP	4	40	4.4
Birth hypoxia	2	20	2.2
Ophthalmia neonatorum	1	10	1.1
Iatrogenic - harmful drops	1	10	1.1
Harmful traditional medicine	1	10	1.1
Surgical complications	1	10	1.1
Subtotal perinatal	10	100	11.1
Postnatal (Childhood) Causes			
Ophthalmia (epidemics), 1 small pox	4	7.2	4.4
Other ocular infections	5	9.1	5.5
Ocular inflammation	2	3.6	2.2
Measles	9	16.4	10
Trachoma	2	3.6	2.2
Vernal Catarrh	2	3.6	2.2
Unidentified ocular cause: cornea 3, secondary glaucoma 1, unidentified 1	5	9.1	5.5
Vascular	1	1.8	1.1
Ocular trauma	10	9.1	11.1
<i>Subtotal</i>	40	72.7	42.2
CNS causes			
Meningitis/encephalitis	6	11	6.6
Intracranial tumours	2	3.6	2.2
Intracranial thrombosis	1	9.1	1.1
Spontaneous subdural haemorrhage	1	9.1	1.1
Cortical blindness	1	9.1	1.1
Trauma - head injury	1	9.1	1.1
Undetermined CNS causes	2	3.6	1.6
Subtotal CNS Causes	14	25.5	15.5
Undetermined	1	1.8	1.1
Subtotal childhood causes	55	100	61
Grand Total	90		100

Table 18-4: Non-genetic Factors

Trauma accounted for 11 cases distributed between the WB (6) and the GS (5) (WB/GS ratio 1.2:1 compared to a population ratio of 1.78:1). M/F ratio was 1.2:1. Of these cases only 1 was from head injury and the rest were ocular including 4 from bomb explosion (Plates 20 to 24).

Blindness ranked very high at 83% of the total with 60% having NLP (n=33). The figure reaches 87% when combining the blind with the svi. Table 18-4 sums up all the conditions in the non-hereditary conditions and Table 18-5 demonstrates the distribution of non-hereditary conditions by region and age.

	West Bank			Gaza Strip			Total Series ^a		
	<16	16+	Total	<16	16+	Total	<16	16+	Total
Prenatal	11	5	16	4	1	5	18	6	24
Perinatal	4	3	7	3	0	3	7	3	10
Postnatal	10	21	31	2	19	21	14	41	55
UD	1	-	-	-	-	-	1	-	-
Total	26	29	55	9	20	29	40	50	90

^a Includes additional 6 from other regions or when the region is uncertain.

Table 18-5: Non-Hereditary Conditions in the WB and GS

18.6 Undetermined Aetiologies (Table 18-6)

In 60 patients it was not possible to ascertain the aetiology. The lack of available family information and pedigree charts in this group was an important factor in this shortfall especially as 55 of these patients were recruited from the out patients at SJOH. Also, the lack of detailed information made it impossible to compile data on the marriage patterns and consanguinity. Conditions of undetermined aetiology formed 13.3% and 15.6% of the pedigree and sibships respectively.

The age groups in the undetermined cases are predominantly under the age of 16 (n=56) and form 9.6% of the total series. The M/F ratio is 2.3:1 in the WB and 2.5:1 in GS. The ratio resembles that of the non-hereditary postnatal cases in the under 16 (Table 18-5). The regional distribution of

patients in this group was; WB 27, GS 16, undetermined WB/GS 16 and Israel 1.

<u>Undetermined Conditions</u>		
Possible genetic aetiology	50	(83.0%)
Possible non genetic aetiology	5	(8.3%)
Prenatal factors (1)		
Postnatal factors (4)		
Either prenatal or genetic	2	(3.3%)
Postnatal or genetic	1	(1.6%)
Undetermined	<u>2</u>	<u>(3.3%)</u>
Total	60	100

Table 18-6: Undetermined conditions

In this group the main site of pathology was the lens in 66.6% (n=40) followed by the whole globe 18% (n=11). The main conditions diagnosed in this group were; congenital cataract (n=40), buphthalmos (n= 9) and others (n=11).