

APPENDIX 2

Family Figures

Ap2.1 Family Information

Prenatal and Postnatal Data in the Total Series

Data were collected from mothers on the prenatal history of all pregnancies including number of miscarriages, abortions and stillbirths. In addition, information was also gathered on the number of live birth and deceased siblings.

Table Ap 2.1a Family data: miscarriages, postnatal mortality, siblings alive, postnatal mortality and deceased affected sibling in the sibships in the survey of childhood onset blindness in the West Bank and Gaza Strip 1985-1987.

	Cohort	Avg.	M:F	MR
Miscarriages				
Total	297	0.82	-	10.6
Hereditary	236	0.83	-	12.5
Non-Hereditary cohort (69 sibship)	54	0.8	-	11
Prenatal mortality				
Total	358	1	-	12.8
Hereditary	290	1	-	13.8
Non-Hereditary	22	0.45	-	12.5
Alive				
Total Live births	2443	6.6	1:1	-
Total gestations	2801	7.6	1:1	-
Alive siblings	2326	-	1:1	-
Postnatal mortality				
Total	320	0.8	0.8:1	12.7
Hereditary cohort	265	0.9	1.25:1	14.7
Non-Hereditary cohort	35	0.7	0.87:1	10
Affected Siblings Mortality				
All	25 (3.5%) ^a	-	0.9:1	10 ^b
Hereditary	37	-	1.25:1	13 ^b

^a Percentage from the total affected. MR: mortality rate.

^b Percentage of deceased affected siblings from the total series.

Table Ap2.1b Gestation and birth rate in total sibships in the survey of childhood onset blindness in the West Bank and Gaza Strip 1985-1987.

	Gestations		Live Births		Living Siblings		
	Nos.	Avg	Nos.	Avg	Nos.	Avg.	M:F
Rod cone dystrophies	472	7.4	390	6	402	3.6	1.06:1
Cone and CRD	268	6.9	247	6.5	236	6	0.86:1
Achromatopsia	137	7.6	130	6.5	121	6.7	0.7:1
Congenital Cataract	359	7.5	343	7	325	7	0.6:1
Congenital Glaucoma	223	6.7	220	6.7	195	6	1.25:1
Microphthalmos	140	7	25	1.25	113	6	1.6:1
Albinism	79	8.8	75	8	72	8	0.8:1
Other Syndromatic	171	7.4	140	6	144	6.3	5:1

CRD: Cone-rod dystrophies; Avg²: Average per sibship.

Ap2.2 Prenatal and Postnatal Mortality in the Clinical Conditions

Ap2.2 Prenatal and Postnatal Mortality in the genetic Conditions in the survey of childhood onset blindness in the West Bank and Gaza Strip 1985-1987.

	Miscarriages			Total Prenatal Mortality			Postnatal Mortality				Affected Siblings Deceased			
	Nos.	Avg	MR	Nos.	Avg	MR	Nos.	Avg	M:F	MR	Nos.	Avg	M:F	MR
RCD	72	1.1	15.3	82	1.3	17.4	59	0.9	1:1	11.3	12	0.2	2:1	18.8
Cone and CRD	22	0.6	8.2	32	0.8	11.9	40	1	0.9:1	14.5	3	0.1	0.5:1	7.7
Cong. Cataract	30	0.65	8.4	34	0.7	9.5	54	1.1	1.9:1	14	1	-	-	2
Congenital Glaucoma	25	0.76	11	28	0.85	12.6	24	0.73	1.25:1	11	6	0.2	0.7	18.2
Achromatopsia	10	0.6	7.3	16	0.9	11.7	17	0.9	1:1	12.3	0	0	0	0
M,phthalmos	25	1.25	18	25	1.25	18	13	0.6	0.9:1	10.3	4	0.2	1:1	20
Albinism	6	0.7	7.6	7	0.7	8.9	13	1.4	1:1	15.3	4	0.4	1:1	44.4
Other Syndromatic	27	1.1	15.8	31	1.3	18	13	0.6	1.2:1	8.3	2	0.09	1:1	8.7

Percentages in **bold Italic**. RCD: Rod-cone dystrophies; CRD: Cone-rod dystrophies; M³phthalia: Micrphthamia; Avgg³: Average per sibship; MR: Mortality rate in %.

Ap2.3 Mortality in Patients in the Total Series

From the total number of 669, 4 patients died during the 2-years period of the study. Two of them died of metastatic retinoblastoma and 2 in their first two years of life from failure to thrive.

The figure gathered from the history on the number of affected patients not examined that died was 25 cases. This together with the four who died during the study, make up a 4.2% mortality rate in the blind population.

Ap2.3 Figures and ratios in sibships of patients with non-hereditary conditions including prenatal and postnatal mortality rates.

	Prenatal	Perinatal	Postnatal	Total
No. of sibships studies	22	7	38	69
Total gestations	173	46	303	535
Average gestations per sibship	7.2	5.7	5.5	6
Total siblings	137	51	248	427
Average siblings per family	6.2	4.2	5.5	6
Total number of patients	24	10	55	90
Sibships with details known	21	8	38	67
Prenatal mortality rate %	10	10	12	12.5
Postnatal mortality rate %	7	19.5	10	10