16

Marriage Patterns and Consanguinity

IK Jalili

16.1 Cohort Size

It was possible to ascertain the pattern of parental marriage in 201 sibships. (Table 16.1)

The symbols used in denoting marriage patterns and the subtypes of cousin marriages are found in the endnote.

Table 16.1 The cohort of families with documented marriage pattern

Cohorts with Documented Marriage Patterns			
	Patients	Sibships	Pedigrees
West Bank	289	201	175
Gaza Strip	259	142	105
Others	9	8	8
Total	557	351	288

16.2 Consanguinity

In the West Bank (WB), consanguineous marriage was found in 80% of the total marriages and included 66% traceable cousin marriage and a much lower figure of 13.4% of untraceable extended family marriages. In the Gaza Strip (GS), the corresponding data were 89.5% for the total consanguineous marriages (traceable, 80.4%; and untraceable 9%. (Table 16.2, Figure 16.1).

Table 16.2 Percentages of consanguinity by aetiology and region

	West Bank			Gaza Strip		
	Cousins	Family	Total	Cousins	Family	Total
Hereditary						
<16	74	7.9	82	83.6	11.4	95
16+	61	21	82	85.7	4.1	90
Subtotal	68.6	13.5	82	84.5	8.2	92.7
Non-Hereditary						
16	47	21	68	66.6	11	77.7
16+	65	9.5	74.5	57	14.2	71.4
Subtotal	56	15	71	60.8	13	74.8

Cons.: Consanguinity

16.3 Consanguinity by Aetiology

There are differences in the proportion of consanguinity between the hereditary and

acquired cases. In the WB, the figures are 82% versus 72% and in the GS 92% versus 74% respectively. (Tables 16.2)

16.4 Temporal Increase of Consanguinity

The rate of consanguineous marriage over the 60 years leading to the study and ending in 1997, by region, are presented in 3 age cohorts; namely the sibships of children <15, the sibships of young adults between 15-30 years and sibships of patients over 35 years of age. Table 16.3, Figure 16.1, 16.2 an the increase in the rate of cousin marriage with a corresponding decrease in the rate of marrying from the same locality /village and unrelated marriages and from acquired condition cohort whicn=h may reflect the trend in the general population.

Table 16.3 Consanguinity in successive generations

Age		Cousins		Family		Total		Same Village		No Relation		Total	
WB	<15	106	***65***	19	***12***	125	***77***	9	***5.6***	28	***17***	162	***100***
	15-30	68	***64***	20	***19***	88	***83***	9	***8.4***	10	***9.3***	107	***100***
	>30	10	***56***	3	***17***	13	***72***	3	***17***	2	***11***	18	***100***
GS	<15	115	***85***	8	***6***	123	***91***	6	***4.4***	6	***4.4***	135	***100***
	15-30	75	***80***	5	***5***	80	***85***	10	***11***	4	***4.3***	94	***100***
	>30	15	***68***	4	***18***	19	***86***	2	***9.1***	1	***4.5***	22	***100***
Both	<15	221	***74***	27	***9***	248	***84***	15	***5.1***	34	***11***	297	***100***
	15-30	143	***71***	25	***12***	168	***84***	19	***9.5***	14	***7***	201	***100***
	>30	25	***68***	7	***19***	32	***86***	3	***8.1***	2	***5.4***	37	***100***

Figure 16.1 Temporal trends of consanguinity by region by age cohort

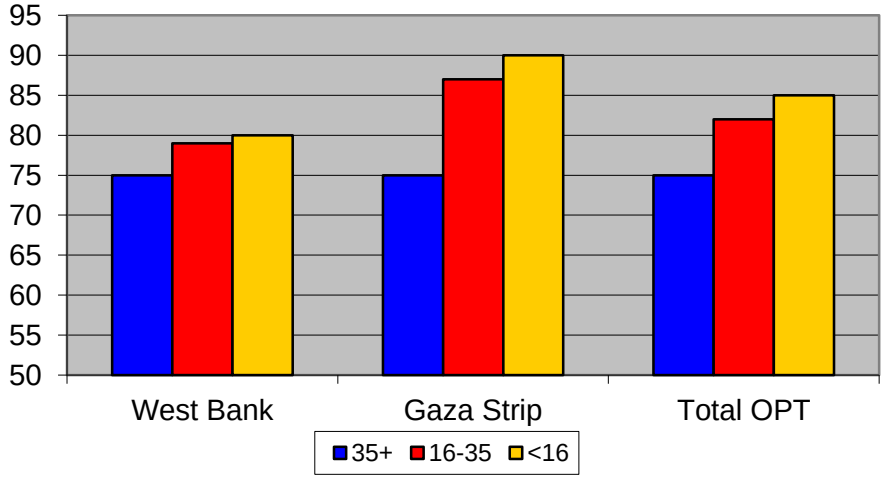
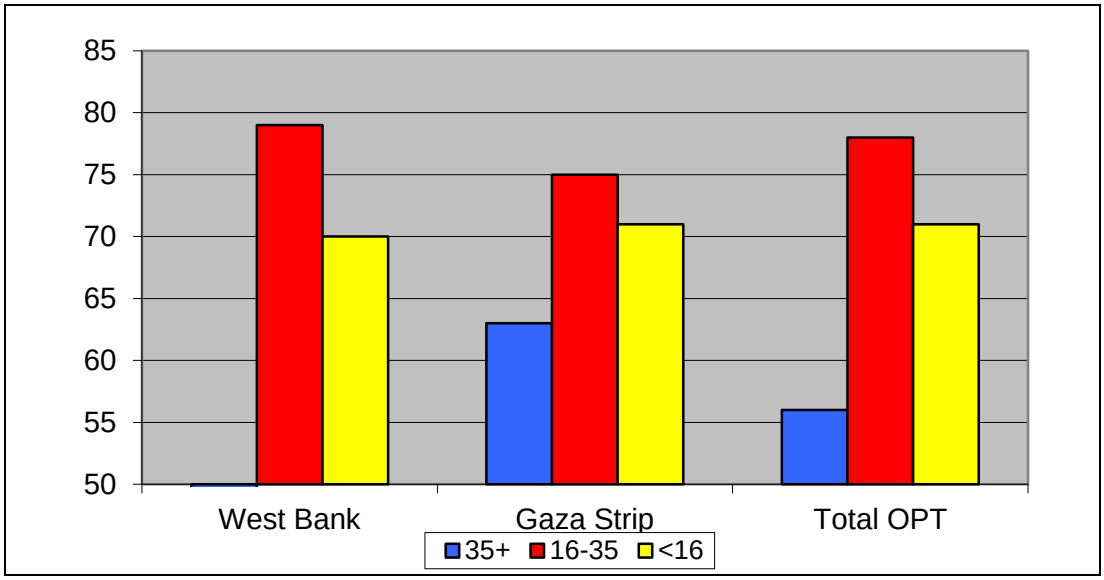


Figure 16.2 Temporal trend in consanguinity in acquired condition



16.5 Subtypes of consanguineous marriage (Social Concepts)

The frequency of the types of first cousin marriages among the Palestinians in the study showed that type A formed 35.4% (n=83 sibships) of all cousin marriages in both regions. This was followed in frequency by type B at 13% (27 sibships) in the WB. In the GS, type C was the second most common marriage at 10.5% of the total (11 sibships). Marriages from the paternal side were 42% (99 sibships) and from the maternal side was 20% (47 sibships was the second most common marriage at 10.5% of the total (11 sibships). Marriages from the paternal side were 42% (99 sibships) and from the maternal side was 20% (47 sibships). (Figure 16.2, Table 16.4)

Figure 16.2 Graphical presentation of the degree (types) of consanguineous marriages

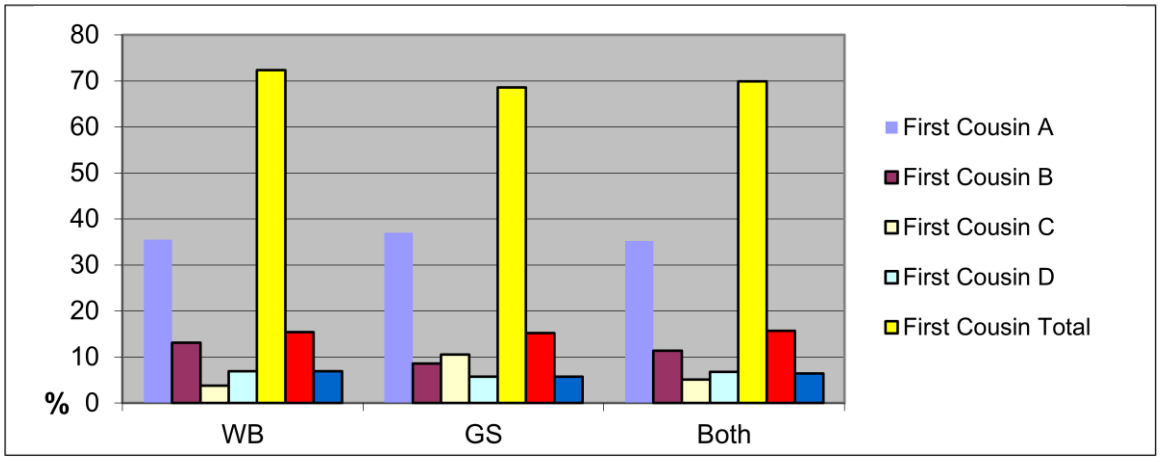


Table 16.4 Subtypes of cousin marriages to reflect social customs

Cousin Marriages	WB		GS		OPT	
First Cousin (type A)	46	***35.4***	37	***35.2***	83	***35.2***
Double first cousin	1	***0.8***	0	***0***	1	***0.4***
First Cousin (type B)	17	***13.1***	9	***8.6***	27	***11.4***
First Cousin (type C)	5	***3.8***	11	***10.5***	12	***5.1***
First Cousin (type D)	9	***6.9***	6	***5.7***	16	***6.8***
First Cousin (either A or C)	3	***2.3***	1	***1.0***	4	***1.7***
First Cousin (either B or D)	4	***3.1***	0	***0***	4	***1.7***
First Cousin (type unknown)	9	***6.9***	8	***7.6***	18	***7.6***
Subtotal first cousin *	94	***72.3***	72	***68.6***	165	***70***
First Cousin once removed	9	***6.9***	6	***5.7***	15	***6.4***
Second Cousin	20	***15.4***	16	***15.2***	37	***15.7***
Second Cousin once removed	3	***2.3***	4	***3.8***	8	***3.4***
Third Cousin	2	***1.5***	5	***4.8***	7	***3.0***
Cousin details unknown	2	***1.5***	2	***1.9***	4	***1.7***
Subtotal	**36**	***27.7***	**33**	***31.4***	**71**	***30***
TOTAL	130	***100***	105	***100***	236	***100***
Paternal Side Marriages	54	***41.5***	49	***46.7***	99	***41.9***
Maternal Side Marriages	30	***23.1***	15	***14.3***	47	***19.9***

* Includes 3 and 4 sibships with double first cousins from the WB and GS.

16.6 Consanguinity in various clinical entities

The highest consanguinity is found in retinal dystrophies, microphthalmos and albinism, making 80% of cases followed by CG (72%).
east are in anterior segment disorders (18.2%) followed by idiopathic nystagmus (28%). (Table 16.5)

Table 16.5 Consanguinity in the common clinical conditions

	Cousins		**Total Cons.**		**Same Location**		**Not Related**		**Total**	
Retina										
Cone Degeneration	6	***100***	6	***100***	0	***0***	0	***0***	6	***100***
Rod-Monochromatism	16	***89***	16	***89***	0	***0***	2	***11***	18	***100***
Cone-rod	21	***78***	23	***85***	2	***7***	2	***7***	27	***100***
Leber's Amaurosis	32	***68***	39	***83***	6	***13***	2	***4***	47	***100***
Rod-Cones	42	***68***	52	***84***	6	***10***	4	***6***	62	***100***
CACR	4	***67***	4	***67***	2	***33***	0	***0***	6	***100***
Macular Degeneration	8	***89***	8	***89***	1	***11***	0	***0***	9	***100***
Vitreo-retinopathies	5	***71***	6	***86***	0	***0***	1	***14***	7	***100***
Albinism	7	***88***	7	***88***	1	***13***	0	***0***	8	***100***
Myopia	10	***83***	10	***83***	1	***8***	1	***8***	12	***100***
Anterior Segment / Whole Globe										
Cong. Glaucoma	27	***75***	32	***89***	3	***8***	1	***3***	36	***100***
Cong. Cataract	30	***65***	35	***76***	4	***9***	7	***15***	46	***100***
Microphthalmos	13	***62***	17	***81***	3	***14***	1	***5***	21	***100***
Colobomas	3	***60***	5	***100***	0	***0***	0	***0***	5	***100***
Syndromatic	12	***55***	13	***59***	3	***14***	6	***27***	22	***100***
Ant. Segment Disorders	3	***50***	3	***50***	1	***17***	2	***33***	6	***100***
Idiopathic Nystagmus	1	***33***	2	***67***	0	***0***	1	***33***	3	***100***
Non-Hereditary										
Acquired Conditions	18	***56***	22	***69***	5	***16***	5	***16***	32	***100***
Optic Nerve conditions	10	***59***	12	***71***	1	***6***	4	***24***	17	***100***

Percentages in ***bold Italic***. Cons.: consanguinity**.**

16.7 Analysis of Marriage Patterns and Consanguinity

This study has also highlighted the high prevalence of consanguinity, and in particular first cousin marriage in this community and the higher consanguinity rate among the families of the blind in comparison with the rest of the population, this is in line with findings in other countries and in particular other Arab countries including neighbouring Jordan. [(1), (2)] (Table 16.6) This difference is noticeable between sibships of the hereditary and acquired conditions, a disparity wider in the <16 cohort being 74% and 47% respectively. (Table 16.2) This is in line with the evidence that higher consanguinity causes an increase in genetic disorders. [(3), (4), (5), (6), (7), (8), (9)] In addition, the higher prenatal and postnatal mortality rates in the genetic series than in the acquired cases (Appendix B) reflects other reports that show the increase in pre-reproductive mortality with inbreeding and the higher under 5 mortality in first cousin marriage. [(10), (11), (12)]

This high consanguinity rate is reflected in the high prevalence of recessive conditions seen in the study which parallels the rate of inbreeding in the community. This is found both in communities where cousin marriage is not the custom as well as in those who practise cousin marriages [**(13), (14), (15), (16)**].

Like the rest of the Arab and Muslim world, first cousin marriages are the commonest type of consanguinity in both regions, although it is higher in the GS, with Type A (son and daughter of two brothers) being the commonest followed by Type B (son and daughter of two sister). Such information is important in any educational campaign especially in view of the common misconception that maternal cousins' marriage is not a relative marriage and does not influence the outcome. Regional variations exist in the pattern of cousin marriage between the WB and GS with a higher percentage of marriages from the paternal side in the Gaza Strip (46.7%) than the WB (41.5%). Conversely, marriage from the mother's side of the family was more common in the WB (23%) than the GS (14.3%).

The study has also shown a temporal increase in consanguinity in the preceding 35 years in both Palestinian regions (Figure 16.2). In the non-hereditary cohorts, however, there is a suggestion of a decline in the consanguinity rate, perhapse this suggests a decline in the consanguinity in general population.

The possibility that the rate of consanguinity, and consequently the prevalence of recessive conditions, is different between the refugees and original town and village settlers could not be confirmed or otherwise in this survey as there was a lack of clarity in the definition of the status of refugees in the collected data.

It was also noted by the author that the presence of blindness in sibships can stigmatise these families, especially when several members are affected, and lead to their isolation, as was the case with the pedigree with CRDAI (Figure 16.4). In this pedigree, individuals descended from the mutant person (who has been identified by the members of the pedigree) are avoided by the remaining extended family. These siblings find no alternative other than taking the risk of marrying from within the family instead of marrying from the extended family or the rest of the village. This can be described as double inbreeding or consanguinity fait accompli.

16.8 Consanguinity Discussion

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. [(11)] 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. [(17)]

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, reaching 80% of marriages in the WB and 89.5% in the GS. These compare with those of the genetic conditions in the Jordanian study (Tables 16.6), which is 79%. This agrees with the common finding of increased morbidity in association with consanguinity (Chapter 9). 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 recessive disorders is the highest recorded among the studies. (Table 16.7) The author believes the degree of accuracy attained in this study is due to the detailed histories taken and pedigree charts drawn up 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.

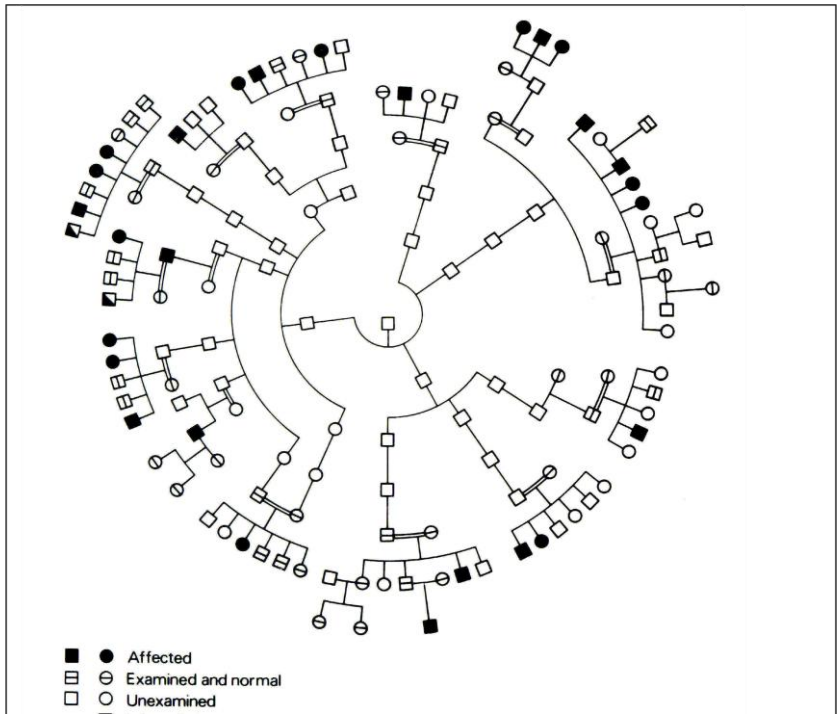
In the MEC the high proportion of genetic disease due to AR conditions (50-65%) has also been caused by the high levels of consanguineous marriage practised which range from 16-55% of all marriages. [18]

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. Many of the children in the study came from very large families with several affected siblings and there was a history of parental consanguinity, usually between first cousins. Table 16.6 compares the results of consanguinity in the West bank and Gaza Strip to some other world figures. (See also Chapter 9 on marriage patterns)

Tables 16.6 Comparison of marriage patterns in the West Bank and Gaza Strip with other Arab and Muslim communities Comparison of Marriage Patterns

	Cousins	Total Family Marriages	Same Locality	Not Related
Genetic Cases				
West Bank	74.2	82.0	6.7	11.2
Gaza Strip	83.6	95.1	4.9	0.0
OPT Total	77.8	87.6	5.9	6.5
Jordan [33]	79	79	-	-
Uzbekistan [43]	33	33	-	-
Bradford [94]	62	62	-	-
Non-Genetic Cases				
West Bank	47.4	68.4	10.3	10.5
Gaza Strip	66.7	77.8	11.1	11.1
OPT Total	54.8	71.0	19.4	9.7
Jordan [33]	33	33	-	-
Uzbekistan [43]	-	-	-	-

Figure 16. 4 Extended pedigree with cone-rod dystrophy and amelogenesis imperfecta (Jalili syndrome) [(19)]



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