

Some Effects on the Offspring of Uncle-Niece Marriage in the Moroccan Jewish Community in Jerusalem

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INTRODUCTION

Considerable data exist on the effects of consanguineous marriage on the offspring of such a marriage (see [1, 2] for review). The investigations almost exclusively deal with first cousins and more distant cousins since marriages of closer than first cousins are rare. Children of incest (brother \times sister, father \times daughter matings) were investigated by Adams and Neel [3] and Carter [4]. Objectively assembled series of offspring of uncle-niece marriages do not seem to exist. It was suggested that such data may contribute to the question of synergism of gene action in man [2], since the coefficient of inbreeding ($F = .125$) of uncle-niece marriage is the highest that permits outcome to be investigated among legal and nonincestuous unions. The relatively high frequency of such marriages among several Jewish communities in Israel made such an investigation possible.

This report will describe a small series of 27 children and their sibships resulting from 27 uncle-niece marriages, and a like number of children whose mothers were not related to their husbands but were as nearly as possible matched to the mothers who were married to their uncles. The mothers in the series and the control mothers were interviewed when pregnant during the years 1966-1968 and were followed up through the Jerusalem Perinatal Study [6].

SUBJECTS AND METHODS

Since 1964, the outcome of all deliveries to residents of West Jerusalem has been recorded and the newborn followed prospectively as part of the Jerusalem Perinatal Study [6]. For a limited period, 1966-1968, it was possible to interview a large sample of the women during pregnancy [7]. At that time, some 88% of all pregnant women attended municipal antenatal clinics and each woman was interviewed shortly after her first attendance. The questionnaire included details of personal and obstetric history, as well as questions about kinship between husband and wife. Eighty percent of the interviews took place during the second trimester of pregnancy. The data were coded and transferred to magnetic tape to be linked later with birth record information. This basic data file was updated periodically with information derived from well-baby clinics, hospital admissions, and other sources.

The original questionnaires of women who stated that they married their uncle were

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reviewed. Cases in which the uncle was only a half uncle or the original questionnaire was missing or inaccurate were discarded. Because of the need for controls, the study group was confined to Jewish immigrants from Morocco, the only large group in Jerusalem with a high (1.6%) frequency of uncle-niece marriage at that time.

For each of the 27 women in the study group, a control married to a nonconsanguineous husband was selected from the data bank, matched by the following criteria:

1. Country of birth: identical (Morocco) for woman and husband.
2. Age at interview: ± 2 years maximum.
3. Degree of religiosity: high degree of observance or not.
4. No. years schooling: ± 3 years maximum.
5. Date of immigration: ± 3 years maximum.
6. Outcome of pregnancy: live birth.

Information on the outcome of all pregnancies of each of the women was retrieved. For the most part this was based on the history but, in addition, the outcome of the present pregnancy and progress of the infant ascertained prospectively. All women were followed to the end of 1971; details of the outcome of subsequent pregnancies, provided that they occurred in Jerusalem, were added to the files.

All births occurred in the hospitals and the infants were examined by a pediatrician soon after birth. All abnormalities were recorded as were abnormalities subsequently reported from well-baby clinics or as a result of hospitalization during the first year of life. These records were linked to the birth data as stored in the data bank.

RESULTS

Of the 27 uncle-niece marriages studied, 14 were maternal uncles and 13 paternal (table 1), indicating that in the Moroccan Jewish community there was no preference as to the type of uncle-niece marriage.

TABLE 1

COMPARISON OF DATA ON OFFSPRING OF STUDY GROUP AND CONTROL GROUP MARRIAGES

SERIAL No.	PROBAND			PREVIOUS PREGNANCIES	
	Year of Birth and Maternal Age	Birth Weight (g)	Sex	N	No. Live Births
Study mother:					
1—M*	1967, 25	3,320	♂	2	2
2—M	1966, 23	3,450	♂	2	2
3—M	1966, 21	3,300	♀	2	1
4—P	1966, 26	3,700	♂	3	2
5—M	1966, 31	4,250	♀	7	6
6—M	1966, 41	3,500	♀	8	6
7—P	1968, 23	3,850	♂	1	1
8—P	1966, 23	2,800	♂	1	1
9—P	1966, 30	2,600	♀	7	6
10—P	1966, 33	4,050	♀	?	?
11—P	1966, 38	3,000	♀	10	7
12—P	1968, 31	3,500	♂	4	3
13—M	1968, 21	3,350	♂	1	1
14—P	1968, 34	3,750	♂	10	9
15—M	1968, 38	3,750	♀	12	10
16—P	1966, 45	2,800	♀	9	9
17—M	1968, 19	3,480	♂	0	0

TABLE 1 (Continued)

SERIAL No.	PROBAND			PREVIOUS PREGNANCIES	
	Year of Birth and Maternal Age	Birth Weight (g)	Sex	N	No. Live Births
18—P	1967, 40	2,770	♀	11	6
19—P	1968, 22	3,580	♀	2	2
20—M	1967, 41	3,260	♀	10	9
21—M	1967, 39	3,400	♀	4	4
22—P	1967, 22	3,700	♂	1	1
23—M	1968, 29	3,120	♂	6	6
24—M	1968, 38	2,500	♂	13	12
25—M	1968, 30	3,550	♀	12	10
26—M	1967, 31	3,700	♂	10	9
27—P	1966, 36	3,320	♀	7	6
Control mother:					
1C	1967, 25	3,250	♀	6	3
2C	1966, 22	3,460	♀	1	1
3C	1967, 20	2,950	♂	1	1
4C	1968, 25	3,150	♂	0	0
5C	1965, 31	3,450	♂	8	7
6C	1967, 41	3,420	♂	7	5
7C	1968, 21	3,740	♂	1	1
8C	1968, 23	3,550	♂	3	3
9C	1967, 31	3,650	♂	5	5
10C	1968, 33	3,800	♂	14	13
11C	1966, 37	4,050	♂	13	12
12C	1967, 31	3,450	♂	5	5
13C	1967, 22	3,350	♂	0	0
14C	1966, 34	2,820	♀	9	5
15C	1968, 38	3,700	♀	10	10
16C	1966, 43	3,270	♀	12	11
17C	1967, 20	3,250	♂	1	1
18C	1967, 40	3,500	♂	10	10
19C	1967, 22	3,350	♂	1	1
20C	1968, 40	3,700	♀	16	13
21C	1967, 38	3,060	♀	6	6
22C	1966, 20	3,100	♂	0	0
23C	1968, 30	3,200	♂	6	5
24C	1968, 38	3,450	♀	10	8
25C	1966, 30	3,300	♀	2	1
26C	1966, 30	4,100	♂	8	7
27C	1966, 36	3,320	♀	13	13

* M denotes marriage to maternal uncle, P to paternal uncle.

The sex distribution of the probands was 14 ♀ : 13 ♂ and that of the control probands, 10 ♀ : 17 ♂. There were no cases of low birth weight either among the probands or control probands, although one proband weighed exactly 2,500 g. The average birth weight of the 67 children born to the uncle-niece unions since 1964 was $3,371 \pm 542$ g and in the control group (81 children), $3,469 \pm 362$ g. This difference is not significant at the 5% level. Malformations detected in the infants of the probands and their controls are listed in table 2. This comparison does not show any difference between the uncle-niece marriage group and the con-

TABLE 2
MALFORMATIONS RECORDED IN OUTCOMES OF STUDY PREGNANCIES

Serial No.	Malformation
Study mother:	
16	Strabismus
19	Umbilical hernia
21	Anteriorly displaced anus
22	Defective motor development
Control mother:	
9C	Pigeon chest
13C	Hydrocele, bilateral
16C	Mongolism, died at 1 month

NOTE.—27 infants in each group.

trol group, although the numbers are small. When considering malformations of previous children as reported by the mother at the time of interview, the number of cases increases as does the period of observation (table 3). Some of the mal-

TABLE 3
MALFORMATIONS REPORTED BY MOTHERS ON CHILDREN BORN BEFORE PROBAND

Serial No.	Malformation
Study mother:	
14	Brother diagnosed with coeliac disease
15	Two siblings mentally retarded
19	Sister with diastasis recti
22	Brother died of congenital heart disease; anomalous drainage of pulmonary vein, hypoplastic left ventricle, partially patent foramen ovale
24	Two siblings deaf
25	Three siblings died of jaundice
Control mother:	
16C	Sister with multiple congenital malformations; died after 15 hr
17C	Brother with congenital heart disease, ventricular septal defect
26C	Brother with inguinal hernia and hydrocele

NOTE.—131 siblings in study group; 134 in control group.

formations reported in these children involve serious diseases that may have genetic cause, such as the case of three siblings who died of jaundice. Ten of the 131 children born before the proband in the uncle-niece marriages (7.6%) were reported to have malformations and only three of the 134 siblings of the control marriages (2.2%). In three of 26 sibships in the uncle-niece marriages, the same

anomaly occurred in two or more siblings (two mentally retarded, two deaf, and three who died of jaundice). No two siblings had the same anomaly in the control group.

There were 22 deaths (16.8%) among the 131 liveborn siblings born before the proband in the uncle-niece marriage group while the figure was only nine deaths (6.7%) among 134 in the control group, a significant difference ($P < .02$; table 4). There were two stillborn infants in each group, and the total number of reported

TABLE 4
SUMMARY OF PREVIOUS PREGNANCIES REPORTED BY THE TWO GROUPS

	PREGNANCIES						
	Live Births	Still- births	Abortions		Total	No. REPORTED MALFORMED*	No. DIED
			Natural	Induced			
Study group	131	2	19	3	155	10	22
Control group	134	2	13	5	154	3	9

NOTE.—Data from 26 uncle-niece marriages and 26 matched controls; study mother no. 10 was excluded because no data were available on previous pregnancies.

* See table 3 for details on malformations.

pregnancies in the two groups was almost identical (155 in the uncle-niece marriage group and 154 in the control group).

At the end of 1971, there were 16 further children born in Jerusalem after the proband to the mothers who were married to their uncles. All were liveborn; one had polycystic kidneys, hydrocele, and bilateral inguinal hernia. There were 26 liveborn children born in the same period to the control group; one had umbilical hernia and another had mild club feet.

DISCUSSION

This first study of 27 uncle-niece marriages is an attempt to bridge the gap between the large scale studies on first-cousin marriages (reviewed in [1, 2]) and the extremely few on incest [3, 4]. The difficulties in study of incest were recently discussed by MacLean and Adams [8]. The finding of suitable controls and the special circumstances involved in incest make both the collection of data and their interpretation extremely difficult. The rarity of uncle-niece marriage in most developed countries of the world is possibly the reason for the absence of reports on objectively assembled series of such matings. Some data on uncle-niece marriages were reported in the middle of the last century by Bemiss [9], but it was pointed out [1] that his method of ascertainment (an appeal to the records of fellow physicians) probably resulted in spuriously elevated consanguinity effects. A considerable number of such marriages are recorded in Sicily [10] and they are rather common in certain parts of India. In contrast to our finding of equal frequency

of paternal and maternal uncles among the uncle-niece marriages in the Jerusalem study, only one paternal uncle as compared to 28 maternal uncles were married to their nieces in Andhra Pradesh because of a very strong tradition [11]. The fact that this mating is permitted by Jewish law and practiced mainly by the North African immigrant communities gave us the opportunity to investigate this group of uncle-niece marriages and select closely matched controls. The probability of inclusion of a family in the study was related to the probability of the mother to be pregnant at the time of the Jerusalem interview survey (a period of 3 years). This results not only in the exclusion of sterile couples but a selection for the more fertile ones and those with fewer early abortions. This bias is the same for both the uncle-niece group and the matched controls.

Extensive studies to date on consanguinity effects have revealed that those who practice consanguineous marriages often differ with respect to socioeconomic status from those who do not [2]. To avoid such socioeconomic differences, the individual matching of each of the 27 women in the study group detailed under the Methods section included number of years of schooling, degree of religiosity, and date of immigration to Israel. These factors are known to be correlated with socioeconomic status. The number of years of schooling of an individual is considered the best indicator of his social status. Another reason for including date of immigration is that medical care in the country of origin may have been on a lower average standard and less available than after immigration, a potential source of bias to our data. To check the success of our matching procedure in selecting controls of equivalent socioeconomic status, an indicator of socioeconomic status not used in the matching process itself was recorded. The number of rooms and the number of persons living in each of the dwellings of both groups are listed in table 5. The average number of persons per room was found to be almost identical in the two groups (3.1 in study group and 3.0 in the control group). It is therefore concluded that differences between the two groups cannot be explained by social differences between them, nor by differences in the availability of medical care. However, the series investigated is small so that the differences found could be due to chance.

The finding of a 10% increase in mortality in the children of uncle-niece couples is in line with the finding of elevated cumulative mortality of approximately 0.75% per percent fetal inbreeding by Schull and Neel [2] in Hirado, Japan. The absence of significant effect of consanguinity upon the frequency of stillborn infants is also in agreement with the Japanese data.

The 3-year period of the Jerusalem interview survey limited the number of recorded uncle-niece marriages available for study in the Moroccan Jewish community to 27. This limited data about the effects of parental uncle-niece marriages may be of some help in the occasional premarital counseling situation when an uncle and niece wish to know the risks involved. However, the numbers are too few to answer the question of synergism of gene action in man. When more or larger series of uncle-niece marriages are reported, perhaps this question will be solved.

TABLE 5
COMPARISON OF NUMBER OF PERSONS AND NUMBER OF ROOMS IN DWELLINGS
OF THE TWO GROUPS

MOTHER'S SERIAL No.	STUDY GROUP		CONTROL GROUP	
	No. Persons	No. Rooms	No. Persons	No. Rooms
1	4	1	5	3
2	4	1	3	3
3	3	2	3	1
4	4	3	3	3
5	8	2	7	2
6	8	2	10	2
7	9	3	3	3
8	3	1	5	1
9	7	2	7	2
10	7	2	11	2
11	9	2	14	3
12	5	2	7	3
13	3	3	2	2
14	10	2	6	1
15	7	3	12	2
16	12	2	10	4
17	2	1	3	1
18	6	2	10	3
19	3	2	3	1
20	10	3	10	3
21	7	2	11	3
22	2	2	2	1
23	9	2	7	2
24	14	3	11	3
25	6	3	3	2
26	8	2	9	3
27	8	3	12	3

SUMMARY

A first attempt to study uncle-niece matings ascertained while the niece was pregnant is reported in the Moroccan Jewish community in Jerusalem. The 27 study families (with 155 previous pregnancies) were compared with individually matched controls of the same socioeconomic status. A 10% increase in mortality at the time of enumeration in the children of uncle-niece couples over the control group was found.

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