

HOW FREQUENT ARE CONSANGUINEOUS MARRIAGES?

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Objective: To determine the frequency of consanguineous marriages in parents and grand parents of all admitted children in a pediatric unit

Design: An observational study.

Methodology: All patients admitted over one year period from January to December 2001 in Unit 1 of Pediatric Department were included. Information was obtained regarding consanguinity of parents and grandparents of the children. Clinically evident inherited disorders were also noted.

Results: Consanguinity was found in 72.7% of marriages. First cousin marriages were seen in 87% while second cousin marriages were seen in 12%. Consanguinity was also seen in the grandparents. It was found in 64.15% of maternal and 60.3% of paternal grandparents.

Conclusion: Cousin-marriages are a norm in the population studied. Its effect on inherited disorders deserves further studies.

KEY WORDS: Consanguinity, Inherited disorder, Social norms.

INTRODUCTION

Consanguineous marriages are frequent in developing countries but their frequency has declined in the developed countries.¹ Studies from various countries and communities in South East Asia show a prevalence of cousin marriages ranging from 20-70% or higher. There is an association of consanguinity with increased incidence of inherited malformations and autosomal recessive disorders.^{1,2} The chances of inheriting biological weaknesses in consanguineous marriages are higher. This exposes the child at a relatively younger age to illnesses and increased chances of succumbing to various inherited disorders.^{4,5} Consanguinity is considered to be the single most important cause of genetically related mortality in developing societies.^{6,7} There has been a decline in consanguineous marriages due to above concerns, in certain countries like Cyprus where there are laws restricting marriages among blood relations while in

China there have been reports of arranged marriages being annulled.⁸ In Turkey there has been a steady decline from one marriage cohort to the next.⁹ Factors attributable may be related to increase in literacy of woman, mobility from rural to urban and an increase in women labour force. Socioeconomic status also has an impact on the pattern of consanguinity. Improvement it showed a decline in consanguinity rates.¹⁰

Pakistan is a country where consanguineous marriages are a societal norm and are widely practiced.² Strengthening family relationships and keeping wealth within the family are some of the reasons for intermarriages.¹¹ Previous studies have shown that in Pakistan consanguinity was seen in 60% of marriages out of which 80% were between first cousins.¹²

Inherited disorders have been found to be twice as common in consanguineous marriages versus non consanguineous marriages.¹³ Relative risks between 1.3 and 2.7 for congenital anomalies had been demonstrated by Khlaf and Khoury for offspring of consanguineous versus non-consanguineous marriages.¹⁴ Considering the population

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of Pakistan and the high fertility rate with the cultural background of consanguineous marriages, the problem of inherited disorders and congenital malformations may be significant. This would add to the high infant mortality rates of the country.¹⁵ It is imperative that more data should be available on consanguineous marriages and the prevalence of inherited disorders to address the issues of antenatal screening and genetic counseling. This study was therefore undertaken to determine the frequency of cousin marriages in the parents and grandparents of children admitted in Paediatric Unit 1 of Civil Hospital Karachi over a period of one year.

METHODOLOGY

All patients admitted in the Paediatric ward over one year period were included in the study. Children were admitted with problems ranging from infection to chronic illnesses and malignancies

Parents were informed about the study and their consent was taken. Information was obtained from the parents of children about the blood relationship between each other and whether it was a first cousin marriage or a second cousin marriage. Similar information was sought about consanguinity in both the maternal and paternal grandparents and also the degree of blood relation. A complete history and physical examination was then performed to establish clinical diagnosis. All patients who had clinical evidence of inherited disorders including autosomal recessive, dominant, X-linked, chromosomal, multifactorial disorders and dysmorphism were documented. Frequencies were calculated and the results were expressed in percentages.

RESULTS

A total of 650 patients were admitted over a one year period. There were 354 (55%) males and 296 (45%) females. Consanguinity among parents was positive in 473 (72.7%) admitted children. Among them 412 (87%) parents were first cousin marriages, while 61(12.0%) were 2nd cousin marriages. Forty two percent were Sindhi and 33% were Balochi. The age ranged from 1 month to 12 years.

Consanguinity was common in maternal and paternal grandparents. It was found in 64.15% of maternal and 60.3% of paternal grandparents.

Seventy seven percent of maternal grandparents were 1st

cousin while 22% were second cousins. Similar results were obtained in paternal grandparents, eighty three (83%) being first cousins while 16.8% were second cousins Sixty patients out of seventy (85.7%) had double consanguinity.

Inherited disorders were found in 70 patients (14.79%) in whom consanguinity was present and 15 patients (8.47%) were with no consanguinity. The male: female ratio was 1.8:1. Autosomal dominant, recessive and chromosomal disorders were found in 5.7%, 17% and 4.2% of patients respectively (Table-I). The lists of disorders are given in tables 2 and 3.

Table 1: Frequency of inherited disorders in the studied children N=70

	NUMBER	PERCENTAGE
Autosomal recessive (AR)	12	17
Autosomal dominant (AD)	4	5.7
Chromosomal	3	4.2
Sex linked	2	2.8
Dysmorphism	9	12.8
Multifactorial (MF)	40	57.1

Table 2: Inherited disorders seen in the studied children with positive consanguinity

INHERITED DISORDERS	NUMBER	PERCENTAGE	CATEGORY
Thalassemia major	6	8.57	AR
Aplastic anemia	6	8.57	AR/AD
Hereditary spherocytosis	1	1.42	AD
Sickle cell anemia	1	1.42	AR
Fanconi's anemia	1	1.42	AR/AD
Wilson's disease	1	1.42	AR
Glycogen storage disorders	1	1.42	AR
Congenital heart diseases	7	10	MF
Congenital adrenal hyperplasia	1	1.42	AR
Epidermolysis bullosa	1	1.42	AD/AR
Insulin dependent Diabetes . mellitus	3	4.28	MF
CP child	9	12.85	AR/AD/MF
Aperts syndrome	1	1.42	MF
Leish-Nyhan syndrome	1	1.42	
Diastrophic Nanism syndrome	1	1.42	AR
Cleft palate	1	1.42	MF
Cruzon syndrome	1	1.42	AD
Trisomy 21	2	2.85	Chromosomal
Ichthiosis	2	2.85	AD/AR
Others	23	32.85	AR/AD/MF
Total	70	100	

* CP = Cerebral palsy, AR = autosomal recessive, AD = autosomal dominant, MF = multifactorial

Table 3: Inherited disorders seen in the studied children with negative consanguinity

	NUMBER	PERCENTAGE	CATEGORY
Thalassemia major	3	20	AR
Congenital heart disease	2	13.33	Multifactorial
Neural tube defect	2	13.33	Multifactorial
CNS malformation	1	6.66	Multifactorial
Insulin dependent Diabetes mellitus	2	13.33	Multifactorial
Dysmorphism	4	26.66	
Muscular dystrophy	1	6.66	X-linked
Total	15	100	

* AR= Autosomal recessive

DISCUSSION

The results indicate that in the admitted patients the frequency of consanguinity was 72.7%. Data from Pakistan Demographic and Health Survey 1990-1991 shows a consanguinity rate of 58.1%.¹⁶ Similar results have been quoted by Bittles.¹⁷ He reported that 50.3% of all marriages in urban Punjab were between 1st and 2nd cousin with prevalence higher in rural areas. The frequency documented in this study was much higher.

First cousin marriages are the most prevalent form of cousin marriage and was also reflected in the present study.¹⁸ Consanguinity rate had shown increase in certain countries. In UAE it had increased from 39% to 50.5%.¹⁹ The reasons stated were primarily socio cultural. Religious beliefs and traditions too may have an effect on the practice of consanguineous marriage, although they were the least cited reason in a study on thalassemia.²⁰

Consanguinity in marriages not only has sociological but also health implications. Such marriages result in transference of recessive gene with a greater chance of congenital malformations and inherited disorders.²⁰ In this study 14.79%, 12.68% and 8.47% of children had inherited disorders with positive single consanguinity, positive double consanguinity and non-consanguineous marriages respectively. Hashmi found an overall highly significant prevalence of congenital malformations in children of related parents it was 40% vs. 26% in non-related parents.²¹ Similar results have been reported from a study done at Ganga Ram hospital Lahore.²² Razaq

found that offsprings of consanguineous marriages had a significantly higher incidence of illnesses (37.1%) than non- consanguineous marriages (29%).²³ A study done at Islamabad showed a strong association of offspring of close relative marriages with cardiovascular diseases compared to controls.²⁴ In the present study, congenital heart disease was most common among the consanguineous off-springs followed by cerebral palsy, thalassemia and aplastic anemia. In the non-consanguineous children, thalassemia was the commonest.

In this study out of 70 children with inherited disorders, 17% children had autosomal recessive disorders, 5.7% had autosomal dominant while 4.2% had chromosomal abnormalities.

Hoodfar and Teehi found the incidence of autosomal recessive disorders to be twice as common in consanguineous unions versus the non consanguineous.² These results can be used for advocacy to discourage cousin marriages especially in families known to have inherited problems as well as for genetic testing of extended families.²⁵

CONCLUSION

Consanguinity is an important issue in the local population. There is a need to conduct community studies on its effect on increase in frequency of inherited disorders. This can be further studied by comparing the frequency with a population without a history of consanguinity for two generations.

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