

Research Article

Role of consanguinity in paediatric neurological disorders

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ABSTRACT

Background: A marriage is said to be consanguineous, where the marriages are solemnized among persons descending from the same stock or common ancestor with close biological relations. The aim of the study was to role of consanguinity in paediatric neurological disorders.

Methods: The cases of the present study were selected from various units of paediatric ward in Velammal medical college hospital, Madurai, Tamilnadu, India, over a period of 2 years between Nov-2013 to oct-2015. This is a prospective observational study of 152 children, out of which 83 children were products of non-consanguineous with neurological disorders and 69 children were products of consanguineous marriage with neurological disorders, confirmed by history and clinical examination and correlated with appropriate investigations.

Results: It was found that out of 152 neurological cases admitted, during the study period, 69 (45.3%) of patients were products of consanguineous marriage and 83 (54.6%) were products of non-consanguineous marriage. Out of 69 cases that were product of consanguineous marriage, 27 cases (39.1%) born of second degree consanguinity and 42 cases (60.8%) were products of third degree consanguinity. Out of the 69 cases, 7 (10.1 %) of them had siblings with similar neurological problems. The prevalence of seizure disorder was 27 (17.7%), developmental delay 13 (8.5%), isolated speech delay 2 (1.3%), hearing impairment 5 (3.2%), mental retardation 11 (7.2%), visual impairment 2 (1.3%), ataxia telangiectasia 2 (1.3%) among the products of consanguineous marriage. The above stated prevalence of neurological disorders being slightly higher than that of the products of non-consanguineous marriage.

Conclusions: It is important to prevent hereditary diseases that are associated with consanguineous marriage through public education.

Keywords: Consanguinity, Recessive genes, Neurological disorders

INTRODUCTION

A marriage is said to be consanguineous, where the marriages are solemnized among persons descending from the same stock or common ancestor with close biological relations. All human societies however primitive or geographically isolated prohibit the mating of first degree relatives. Marriage between relatives less closely than siblings or parents and offspring are not necessarily outlawed, but the dividing line between legal and illegal is hazy and varies between countries. In about one half of USA, uncle-niece and aunt -nephew (second degree) and first cousins (third degree) mating are

forbidden by law. In 1981, in China marriage between first cousins or closer were also prohibited. On the other hand, in India marriage between uncle and niece is legal. It has been reported that in Bangalore and Mysore, two major cities of Karnataka state of south India, 21% of Hindu marriages were uncle – niece unions. The detrimental health effects associated with consanguinity are caused by the expression of recessive genes inherited from common ancestors.¹

Consanguinity means the amount of shared identical DNA. Globally, the most common form of consanguineous union contracted is between first cousins,

when the spouses' share 1/8th of their genes inherited from a common ancestor, where the offspring of consanguineous relationships are at a greater risk of certain genetic disorders. As relatives share a proportion of their genes, it is much more likely that related parents will be carriers of an autosomal recessive gene and their children are at a higher risk of an autosomal recessive disorder, where extent to which the risk increases depends on the degree of genetic relationship between the parents. Increased risk of epilepsy at least by 2.2 folds has been reported after familial marriages in a study from Iran. A recent study in Qatar showed that mental retardation, epilepsy were significantly more common in offspring of consanguineous couples.²

It is estimated that globally at least 20% of the human population live in communities with a preference for consanguineous marriage and that at least 8.5% of children have consanguineous parents.³ African populations, Malians in particular have a high rate of intra ethnic and consanguineous marriage resulting in increased prevalence of autosomal recessive diseases.⁴ In Saudi Arabia, reports showed high rates of consanguineous marriage across regions, which was associated with mental retardation, neural tube defects and other hereditary neurological diseases.⁵ Incidence of consanguinity reported in India is 5-60% and uncle – niece and first cousins union are the more commonly occurring relationships in Indian population.⁶ Researchers declared that consanguinity is the cause of 70% of hearing impairment and deafness. There was significant relation between consanguineous marriage and more than one disabled children in the family as, 77% persons who had more than one disabled child had consanguineous marriage.⁷ Consanguineous marriages are traditionally favored in most of the eastern Mediterranean region, South Asia and African countries.⁸ Consanguinity is a well-known risk factor for genetic disorders including diseases and syndromes that present with intellectual and developmental disabilities.⁹ In both low and high income countries, elevated levels of global developmental delay and mild and severe intellectual and developmental disability have been consistently associated with parental consanguinity.¹⁰

METHODS

This study was undertaken in the department of paediatrics, in Velammal medical college, Madurai, Tamilnadu, India. It was a prospective observational study conducted over a period of 24 months from Nov 2013-Oct 2015. Informed consent was taken from all the patients included in this study. A total of 152 children less than 15 years of age, both male and female with neurological disorders were included in this study. Out of which 83 cases, were products of non-consanguineous marriage. The remaining 69 children, who were products of consanguineous marriage with neurological disorders, were further evaluated.

Children above 15 years of age with neurological problems and patients who were products of consanguineous marriage without neurological abnormality were excluded from this study.

All the patients who were included in this study were assessed by detailed history regarding the neurological disorder, developmental milestones, degree of consanguinity among the parents, siblings affected if any, with similar neurological problems in their family. Residence of the patient was also noted. Detailed neurological examination, estimation of developmental quotient, intelligence quotient was noted. Complete blood counts and urine analysis were done in all cases. Appropriate investigations like neuroimaging, EEG, vision and hearing assessment, CSF analysis were carried out as per case merit. Treatment and observations were done for all cases. Counseling was given for all the consanguineous couples with affected children regarding prevention and perinatal screening for hereditary disorders.

RESULTS

Out of 152 neurological cases admitted in the paediatric ward, during the study period, with neurological disorders, 69 (45.3%) of patients were products of consanguineous marriage, 83 (54.6%) of patients were products of non- consanguineous marriage.

Table 1: Distribution of the degree of consanguinity among the products of consanguineous marriage.

Degree of consanguinity	Number (Out of 69 cases studied)	Percentage
First degree	Nil	Nil
Second degree	27	39.1 %
Third degree	42	60.8 %

Out of the 69 cases studied 27 (39.1 %) were born of second degree consanguineous marriage and 42 (60.8%) were born of third degree consanguinity (Table 1).

Table 2: Number and percentage of siblings affected among the cases studied.

Cases studied	No. of siblings affected	Percentage
Product of consanguineous marriage (total 69 cases)	7	10.1%
Product of non-consanguineous marriage (total 83 cases)	NIL	NIL

It was found that out of the 69 cases, who were products of consanguineous marriage, with neurological problems, 7 (10.1 %) of them had siblings with similar neurological problems. In contrast, out of the 83 cases, who were

products of non-consanguineous with neurological problems, none of them had any siblings with similar problems in the family (Table 2).

Table 3: Distribution of various neurological disorders among cases studied.

Neurological disorder	Product of consanguineous marriage. No./% out of total 152 cases	Product of non-consanguineous marriage. No./% out of total 152 cases	Total (152 cases)
Seizure disorder	27(17.7%)	21 (13.8%)	48
Febrile seizure	7(4.6%)	42 (27.6%)	49
Development delay (Including cerebral palsy)	13 (8.5%)	9(5.9%)	22
Isolated speech delay	2(1.3%)	1 (0.6%)	3
Mental retardation	11 (7.2%)	7 (4.6%)	18
Hearing impairment	5(3.2%)	3 (1.9%)	8
Visual impairment (congenital blindness)	2(1.3%)	Nil	2
Ataxia telangiectasia	2 (1.3%)	Nil	2

It was seen that the prevalence of seizure disorder was 27 (17.7%), developmental delay 13 (8.5%), isolated speech delay 2 (1.3%), mental retardation 11 (7.2%), hearing impairment 5 (3.2%), visual impairment 2 (1.3%) and ataxia telangiectasia 2 (1.3%) among the product of consanguineous marriage (Table 3).

DISCUSSION

This study examined the role of consanguinity in neurological disorders. It was carried out in southern part of Tamilnadu, India, where the prevalence of consanguinity is very high.^{1,18}

The present study observed that products of consanguineous marriage have higher incidence of neurological disorders such as, seizure disorders, isolated speech delay, developmental delay, visual and hearing impairment, mental retardation and ataxia telangiectasia compared to off springs of non-consanguineous union. Similar results were reported by many other authors in various studies all over the world.^{2,6,9-14}

Our study showed a higher incidence of third degree consanguinity 42 cases (60.8%) among couples of affected offspring's than second degree consanguinity 27 cases (39.1%). Similar findings were observed by Nathen et al, in which it was revealed that majority of consanguineous marriage were between first cousins.^{15,16}

It was observed that 7 (10.1%) of the affected children among the consanguineous marriage had siblings with similar neurological problems. Similar results were reported in a study conducted by Mosangebi et al, in

which a total of 5 cases (9.3%) with consanguineous group had a history of affected sibling.¹⁷ According to Anita et al, consanguineous couple supporting one are almost 13 times more likely to give birth to another affected child as compared to non- consanguineous couples.¹⁸

CONCLUSION

It is important to prevent hereditary diseases that are associated with consanguineous unions. Public education has to be undertaken at school level during adolescence to instill the biological risk of close marriage.

It would be particularly advisable to avoid consanguineous marriage in families, where already a child with an autosomal recessive disorder has been born. Premarital and preconceptional testing and counseling for common and rare disorders that are present in high risk inbred families also help in reducing the incidence of inherited disorders in many populations.

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