

Introduction

Consanguineous marriage is defined as a union between two individuals who are related as second cousins or closer. This includes unions termed first cousins, first cousins once removed and second cousins (*Hamamy et al., 2011*).

The rate of consanguineous marriage varies in different countries and is usually associated with some demographic features, as religion, educational level, socio-economic status, location and size of the area, isolation of population, consanguinity in parents' marriages, responders' attitude towards consanguineous marriage, and living in rural or urban area (*Sueyoshi and Ohtsuka, 2003*). Consanguineous marriage remains common in many parts of the world and has been reported in various communities. It is reported as 1% in United States, Russia, Australia, parts of Latin America and Europe. Meanwhile, 1-10% are reported in China, Latin America, North India, Japan, South Europe and Canada. There is a much higher prevalence (10-50%) in Arab countries, Turkey, Iran, Pakistan, Afghanistan, South India. This practice continued in some of the communities who settled the West such as the Pakistani community in the UK (*Gunaid et al., 2004*).

Although the frequency of consanguineous marriage is generally declining, most Middle Eastern Arabs still have a custom of preferring consanguineous marriage particularly among first cousins (*Shawky et al., 2011*).

In the Arab countries, consanguinity has been reported with the highest frequency in Saudi Arabia where it reaches 80% of marriages in certain parts of the Kingdom. Average figures ranges between 59% among the Iraqis, 40% among the Palestinians, 44% among the Yemenis in Sanaa', 49-58% among the Jordanians and 40-54% in the UAE (*Zlotogora et al., 2000*).

Consanguinity is less common in North African Arab countries where it was reported to be 29% in Egypt. However in another study on the Nubian population in southern Egypt the figures ranged between 41.5-45.5%.The highest rates of such marriages have been reported in rural areas, among individuals with low educational levels, and among the poorest (*Sueyoshi and Ohtsuka, 2003*).

It has been a long standing social habit among Egyptians. The etiology of this high degree of consanguinity is nearly the same in all Arab countries. It includes maintenance of family structure, stability, durability as well as keeping property. It has financial advantages relating to ease of marital arrangements. It will also strengthen family ties and build closer relationship between the wife and her in-laws (*Jorde, 2001*).

There are many adverse health effects of consanguineous marriage, it has a greater risk not only of

producing offspring which are homozygous for a deleterious recessive gene, but also individuals with increased susceptibility for polygenic or multifactorial disease, sterility, still births, spontaneous abortions, child death, infant mortality. There is no association of consanguinity with autosomal dominant, X-linked, or chromosomal disorders (such as Down syndrome). Consanguinity increases the risk of expression of autosomal recessive conditions in the offspring (***Bittles, 2001***). Consanguinity is reported to be associated with higher proportions of schizophrenia, several congenital heart defects such as septal defects (atrial, ventricular and atrio-ventricular). Also reported disorders include pulmonary stenosis and atresia, cystic fibrosis, nephropathic cystinosis, spinal muscular atrophy, albinism, hearing disorders, and central nervous system anomalies, congenital anomalies, physical handicap, mental retardation and malignancies. Higher incidence of phenylketonuria, and familial Mediterranean fever have been documented in cousin marriage (***Becker et al., 2002***).

Kafr aburaqaba is a rural area with population preference of consanguineous marriage due to cultural and social habits. There is no previous studies regarding prevalence of consanguineous marriage or the child health related problems associated with it.

Research Hypothesis:

It is hypothesized that the frequency of consanguineous marriage is high in Kafr Abu Raqabah, and consequently high frequency of health-related problems among children.

Research Questions:

The current study will answer the following questions:

- 1- What is the prevalence of consanguineous marriage in Kafr Abu Raqabah?
- 2- What are the frequencies of different health-related problems in children associated with the consanguinity?

Aim of the Work

Long term Objective:

To improve the quality of life of Egyptian population.

Specific Objectives:

- To determine the prevalence of consanguineous marriage in Kafr Abu Raqabah, Ashmone Al Minufiyah Governorates.
- To identify the frequencies of various birth outcomes and child health-related problems due to consanguineous marriage in children e.g. abortion, stillbirths, congenital malformations, genetic disorders, diseases with autosomal recessive inheritance, developmental disorders, congenital heart diseases, hearing problems.

Chapter (1): Consanguineous Marriage

Linguistically, consanguinity is a term that is derived from two Latin words "*con*" meaning common, or of the same and "*sanguineus*" meaning blood, referring to a relationship between two people who share a common blood, Consanguineous Unions (CU) are generally common in the developing world (*Riaz et al., 2016*).

Consanguinity means marriage between two individuals descended from the same ancestor and in practice, this ancestor is usually taken to be no more remote than a great-great grandparent. The genetic definition indicates unions contracted between persons biologically related as 2nd cousins or closer ($F \geq 0.0156$). The coefficient of inbreeding (F), which is defined as the probability that a child inherits two identical copies of an allele from one or more common ancestors (*Shawky et al., 2011*)

A cousin is a relative with whom a person shares one or more common ancestors. Generally, cousins are two or more generations away from any common ancestor, thus distinguishing a cousin from an ancestor, descendant, sibling, aunt, uncle, niece, or nephew. However, in common practice, "cousin" normally specifically means "first cousin" (*Jabeen et al., 2014*).

The ordinals in the terms "first cousins", "second cousins", "third cousins", refer to the number of generations to one's closest common ancestor. The number of "G" words used to describe this ancestor will determine how close the relationship is. For example, having "Great-Great-Grandparents" in common would be third cousins (*Ahmad et al., 2016*).

First cousins

The children of two siblings, First cousins share grandparents. First cousins once removed, who descend from a common ancestor but differ in the number of generations. While double first cousins whom all grandparents are shared (*Jabeen et al., 2014*).

Second cousins

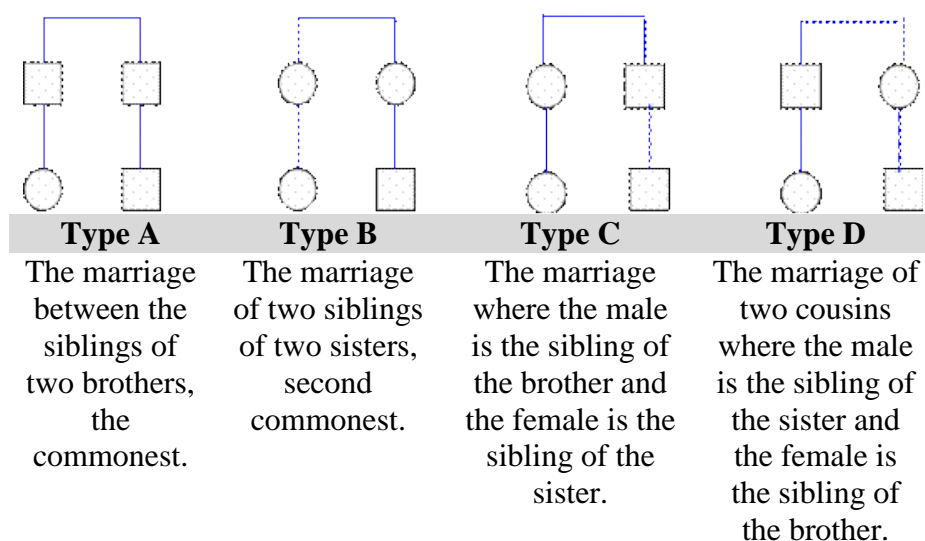
The children of first cousins, second cousins share great grandparents.

Third cousins

The grandchildren of two first cousins, also the children of two second cousins, third cousins share great-great-grandparents. Globally, the most common form of consanguineous union contracted is between first cousins, in which the spouse share 1/8 of their genes inherited from a common ancestor and so their progeny are homozygous

at 1/16 of all loci. This is expressed as the co-efficient of inbreeding (F) and for first cousin offspring (*Shawky et al., 2011*).

The first cousin marriage can be classified to 4 types according to the relationship of the parents and according to the sex of the couple's parents who are sibs as follows (*Naibkhil and Chitkara, 2016*):



1 st degree: Parent/child Brother/sister	share ½ of their genes
2 nd degree: Brother/half-sister Uncle/niece Double 1st cousins	share ¼ of their genes
3 rd degree: First cousins Half uncle/niece	share ⅛ of their genes

Table (1): Consanguinity and genetic inheritance (*Raz et al., 2004*)

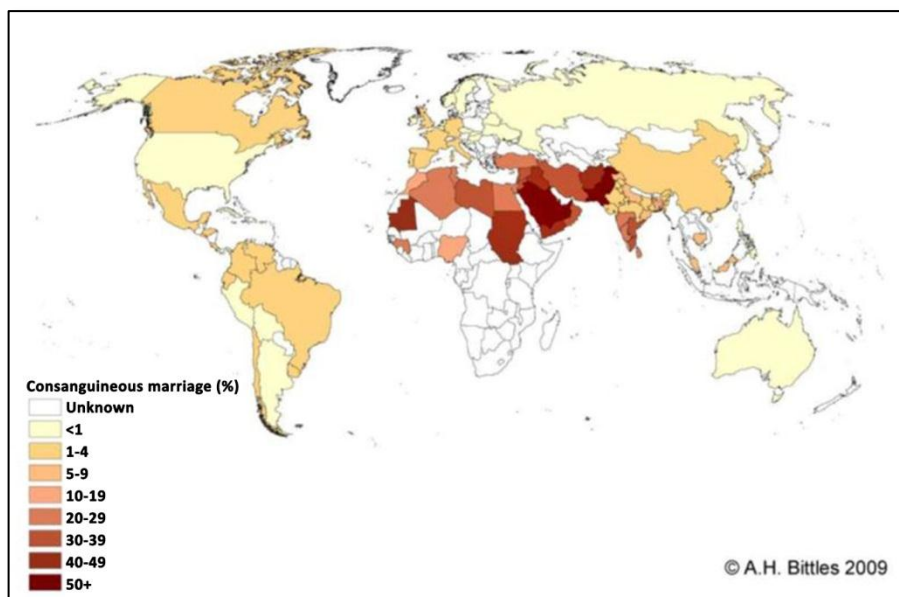


Figure (i): Global distribution of consanguineous marriages. The numbers are lower-bound estimates of the overall global prevalence of consanguineous marriage (*Saggar and Bittles, 2008*).

Global consanguinity trends

- Less than 1%: United States, Russia, Australia, parts of Latin America and Europe.
- 1-10%: China, Latin America, North India, Japan, South Europe and Canada.
- 10-50%: Arab countries, Turkey, Iran, Pakistan, Afghanistan, South India.
- Unknown: Parts of South-East Asia, most Africa.

It is estimated that one billion of the current global population live in communities with a preference for consanguineous marriage.

Prevalence of consanguineous marriages increased in Western countries. Rates in migrant populations are sometimes even higher than in the country of origin. In Japan the rate of consanguineous marriage has been declining and was found to be 8 times lower in younger individuals than the oldest individuals (*Ismail, 2005*).

Middle East, Northern Africa, and South Asia are regions that have historically and culturally had a high rate of consanguineous unions recent studies have shown that 20 % to 50 % of marriages in Arab countries are between relatives (*Halim et al., 2013*).

The lowest rates of consanguinity are found in Western Europe and North America, as less than 1% of marriages are consanguineous. In some parts of Southern Europe, South America and Japan, where the rate ranges 1–10% of marriages are consanguineous, depending on local geography and social customs. A recent decline in the prevalence of consanguineous marriage has been reported in Middle Eastern countries such as Jordan and Saudi Arabia, but increases have been reported in the neighboring Arab states of Qatar and the United Arab Emirates (*Saggar and Bittles, 2008*).

Little change appears to have occurred in the prevalence of consanguinity during the latter half of the twentieth century in the major South Asian countries of India, Pakistan and Iran (*Saggar and Bittles, 2008*).

In populations of North Africa, West Asia and South India, consanguineous marriages are culturally and socially favored and constitute 20-50% of all marriages, with first cousins unions accounting for almost one-third of all marriages. The prevalence of consanguinity and rates of first cousin marriage vary widely within and between populations and communities, depending on ethnicity, religion, culture and geography. Consanguineous marriages are also practiced among emigrant communities from highly consanguineous countries and regions, such as Pakistan, Turkey, North Africa and Lebanon, now resident in Europe, North America and Australia (*Hamamy et al., 2011*).

Marriage between first cousins is a longstanding tradition in the Gulf States, particularly between the children of brothers. Though consanguinity has been rare in Western societies since the early twentieth century, most Western countries are home to large migrant communities which traditionally have contracted consanguineous unions. Evidence points to continued preference for consanguinity in their newly adopted countries in these communities (*Saggar and Bittles, 2008*).

In highly consanguineous populations, families with complex history of consanguineous marriage are resulting in higher inbreeding coefficients. Reports on consanguinity

rates may include marriages between first, second and third cousins or more remote related persons. Although this discrepancy changes the total consanguinity rate, the lower coefficients of inbreeding in more remote unions limit a marked change of the mean inbreeding coefficient (α). The two parameters best used to compare consanguinity rates among different populations are the mean inbreeding coefficient and the rates of marriages between first cousins (*Hamamy et al., 2011*).

Prevalence of consanguinity in the Arab countries:

The rate of consanguineous marriage varies in different countries and is usually associated with some demographic features, as religion, educational level, socio-economic status, location and size of the area, isolation of population and living in rural or urban area (*Shawky et al., 2011*).

Consanguineous marriage has been reported with the highest frequency in Saudi Arabia as it reaches 80% of marriages in certain countries like some regions in Saudi Arabia. Middle East Countries are ranged between 59% among the Iraqis, 40% among the Palestinians, 44% among the Yemenis in Sanaa', 49-58% among the Jordanians and about 40-54% in the UAE (*Ismail, 2005*).

Consanguinity is less common in North African Arab countries where it was reported to be 29% in Egypt; however, in another study on the Nubian population in southern Egypt the figures ranged between 41.5-45.5% (*Shawky et al., 2011*).

Consanguineous marriage in Egypt:

In contrast to the recent decline in the prevalence of consanguineous marriage that was reported in Middle Eastern countries such as Jordan and Saudi Arabia, an increases have been reported in other Arab countries as Qatar and United Arab Emirates which still have a custom of preferring consanguineous marriages (*Shawky et al., 2011*).

It is still prevalent in Egypt especially in rural areas (about 45% of all marriages) where marriage of non-educated and unemployed persons is practiced. The prevalence of consanguineous marriages in Egypt is still high (35.3%), however this frequency varies by region. It was significantly higher in Upper Egypt Sohag (42.2%) than in Lower Egypt Cairo (36.1%) (*Mansour et al., 2010*).

Factors affecting consanguineous marriage:

Arabs and some other communities have preferred consanguineous marriage because its significant social and economic advantages outweigh the disadvantages (*Alharbi et al., 2015*).

There are a number of factors which have direct or indirect impact on the rate of consanguinity. These factors include cultural ties, socioeconomic conditions, family structure, and educational level of parents 'jobs, religion, and geography. Generally the highest rates of such marriages have been reported in rural areas, among individuals with low educational levels, and among the poorest (*Joseph et al., 2015*).

1- Social and cultural factors:

The main causes given for the preference of consanguineous marriages are primarily social. In countries with high consanguinity rates, previous sociological studies indicate that consanguineous marriage could enforce the couples' stability due to higher compatibility between husband and wife who have the same social relationships after marriage as before marriage (*Warsy et al., 2014*).

Consanguineous marriage may be more favorable for the women's status, including the wife's better relationship with her in-laws who could support her in time of need. It is believed that consanguinity strengthens family ties and enforces family solidarity, with cousin marriage providing excellent opportunities for the transmission of cultural values and cultural continuity. Wife's parents prefer to have their daughter living near them and to enjoy the presence of their grandchildren (*Hamamy et al., 2011*).

2- Financial factors:

It is generally accepted that consanguineous marriage strengthens family ties and promotes family stability, simplifies pre-marital preparation, offers a greater compatibility between the spouses and other family members (*Bernadette and Amra, 2014*). There is a general belief that marrying within the family reduces the possibilities of hidden uncertainties in health and financial issues. Premarital negotiations regarding financial matters of marriage are more easily conducted and sometimes less costly (*Hamamy et al., 2011*).

3- Geographical factors:

In communities with increased number of individuals who migrated from their countries to more developed countries there is significant cultural differences faced by the people who migrate to the developed countries. Many of the younger people who, under the influence of the family agree to arranged marriages, in their country of origin, when they migrate to other countries, the influence of family is lessened and the determination to find a partner of their own choice is elevated, hence the effect on the prevalence of consanguinity. Conversely, immigration could also increase consanguinity prevalence, as people feel more confident and sure with people from their own families (*Arjumand et al., 2014*).