

Case Report

Premarital Screening Programs in the Middle East, from a Human Right's Perspective

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ABSTRACT

Premarital screening aims primarily to give couples (who are planning to get married soon) medical consultation on the odds of transmitting genetic diseases to their children. Given that consanguineous marriages are so common in the Middle East and children of consanguineous marriages are at increased risk for genetic diseases and congenital malformations, and due to its impacts on public health, it is important to provide health education on consanguinity at individual, family and community levels delivered mainly by the state's primary health care staff. It is also important to provide pre-marriage and pre-pregnancy genetic diagnosis and counseling. The Availability, Accessibility, Acceptability and Quality (AAAQ framework) of specialized facilities/centers for pre-marriage and pre-pregnancy genetic counseling and testing services should be sufficient to implement the Right to Health in this

regard. And human rights principles shall guide the whole aspects of premarital screening program to include planning, design, execution, monitoring and evaluation. Among those principles are also non-discrimination, participation, inclusion, and accountability. All parties should be involved in the premarital screening program- politicians (to enact laws), religious leaders (to reconsider abortion in case the fetus afflicted with grave congenital malformation), employees in primary health care, researchers, doctors, geneticists, disease prevention and control and social services, subject-matter experts, mother and child health care, media, and activists in NGOs and voluntary organizations.

Keywords: Premarital screening; Middle East; Consanguineous marriages; Sickle cell anemia; hepatitis

Introduction

Premarital screening

Premarital screening is defined as testing couples who are planning to get married soon for common genetic blood disorders (mainly hemoglobinopathies, e.g. thalassemia and sickle cell anemia) and infectious diseases (e.g. hepatitis B, hepatitis C, and HIV/AIDS). The premarital screening aims to give medical consultation on the odds of transmitting the abovementioned diseases to the other partner/spouse or children and to provide partners/spouses with options that help them plan for healthy family. The premarital screening reduces the spread of the abovementioned diseases and reduces the financial burdens of their treatments as well. It reduces the burden on the state's health facilities and blood banks. The screening would avoid any future's social and psychological problems of families. It helps those who seek such medical check-ups feel at ease; premarital screening raises awareness about healthy and sound marriages (Premarital Screening, 2014, April 08). Premarital check-ups may include also testing for syphilis, gonorrhea, and other sexual-transmitted diseases, blood grouping, resus factor, seminal fluid analysis, FSH, prolactin, testosterone, estrogen hormones, among others.

Cyprus was among the first countries that implemented the mandatory premarital testing for β -Thalassaemia in 1973. The program was recently conducted in several countries in the Middle East to include Iran (1997), Saudi Arabia (2004), and United Arab Emirates (2011) [1].

The effectiveness of premarital testing for β -Thalassaemia

in the Middle East and found that this testing program was no success in discouraging consanguineous marriages although it helped reduce affected births in countries where prenatal screening is offered, and therapeutic abortion is considered (e.g. Turkey, Iran) [2].

Consanguineous marriages in the Middle East

Consanguineous marriages or relative marriages (and more specifically first-cousin marriages) are so common in the Middle East and North Africa (MENA) region [2]. Consanguineous marriages constitute 42-67 percent of all marriages in the Kingdom of Saudi Arabia, 54 percent in Qatar, 40-54 percent in the United Arab Emirates, 29-64 percent in Jordan, 21-33 percent in Egypt, 44-63 percent in Sudan, and 40-45 percent in Yemen [3].

Children of consanguineous marriages are at increased risk for genetic-recessive diseases (e.g. thalassemia, sickle cell anemia and cystic fibrosis) and congenital malformations (2.5 times higher than the rate among the children of nonconsanguineous parents) due to the expression of autosomal recessive gene mutations inherited from a common ancestor. The closer the biological relationship between mother and father, the higher is the likelihood that their children will get identical copies of one or more harmful recessive genes [4,5]. Offspring of consanguineous parents have higher neonatal, post-neonatal, and child mortality than those of nonconsanguineous parents [6]. In addition, consanguineous unions are more likely to result in inborn error of metabolism diseases (for example mucopolysaccharidosis, phenylketonuria), multifactorial

disorders (mental retardation, bronchial asthma, diabetes and epilepsy); and sensorineural deafness [7].

The Global Report on Birth Defects, issued by March of Dimes Birth Defects Foundation, New York, 2006, ranked Sudan first (82.0 birth defects per 1,000 live births), Saudi Arabia second, Occupied Palestinian Territory fifth, the United Arab Emirates sixth, Iraq eighth, Kuwait ninth, Oman eleventh, Syria twelfth, Qatar sixteenth, Bahrain seventeenth, Jordan eighteenth, Libya nineteenth, Tunisia twentieth, Morocco twenty-first, and Yemen twenty-second, out of 193 countries with children born with birth defects. The lowest rate is in France with 39.7 per 1,000 live births [8].

Thalassemia

Thalassemia is an inherited blood disease that causes the body to produce lower-than-normal red blood cells and below-than-normal hemoglobin levels (hemoglobin is an oxygen-carrying protein); thalassemic patients can have mild or severe anemia. The beta form of thalassemia (β -thalassemia) is prevalent in the Middle East mainly in the Mediterranean countries with reported carrier rates of 1-11 % for β -thalassemia, 1-58 % for alpha form of thalassemia (α -thalassemia) in the Arab World's population [10].

Sickle cell anemia

Sickle cell anemia is another inherited blood disease; it is common in the Middle East too with rates of sickle cell trait (sickle cell carrier) in the Arab World's population range from 0.3 to 30%. Hamamy and Al-Allawi, Alhamdan et al. [10,11] showed that 4.2 percent of Saudis are carriers of sickle cell anemia (sickle cell trait) and 0.26 percent of them have the disease. The Al-Ahsa Region (in the Eastern Province of Saudi Arabia) has the highest prevalence: 16.89 percent of its population carry the detrimental gene and 1.2 percent of them have the sickle cell disease. Due to the presence of the abnormal hemoglobin S (HbS), red blood cells sickle (i.e. become sickle- or crescent-shaped) upon deoxygenation, leading to health problems like pain (sickle-shaped red blood cells obstruct capillary blood vessels and prevent blood flow to the target organ resulting in ischemic pain), anemia (due to hemolysis of red blood cells), stroke, swelling in the hand and feet, etc.

Hepatitis B

A viral infection that attacks the liver causing acute and chronic illnesses; it transmits through body fluids (blood, saliva, seminal and vaginal fluids). So, sexual transmission of the disease is possible. World Health Organization estimated that 2-5 percent of Middle East populations are chronically infected with hepatitis B. There is no specific therapy for hepatitis B so far; the infection can be prevented by vaccination/immunization; if either of spouses or partners is tested positive for the disease then the other spouse/partner should take the vaccine (usually given as three shots) before they consummate their marriages. Chronic hepatitis B puts patients at increased risk of death from cirrhosis and hepatocellular carcinoma-liver cancer (Hepatitis B, July 2016). Around 10 percent of Egyptians aged 1-59 had

been exposed to the hepatitis B virus at some point; 1% of them have an active hepatitis B infection (Ministry of Health and Population [Egypt] et al., 2015).

Hepatitis C

Hepatitis C is another viral infection that attacks the liver; it is a major cause of liver cirrhosis and liver cancer, and main indication of liver transplantation. It is a prevalent disease in the Middle East. The prevalence of Hepatitis C in Egypt reaches to 15 percent – the highest in the world. It is a blood-borne disease – transmitted through blood and blood product, unsterilized needles and instruments for tattooing and piercing, and sexual exposure as well [12]. Eplusa, a breakthrough drug for hepatitis C has been developed recently by Gilead Sciences Inc. and approved by Food and Drug Administration (FDA) in June 2016; this highly effective drug is taken for 12 weeks; and the course of treatment costs around USD 75,000 [13,14].

HIV/AIDS

HIV/AIDS is a blood-borne and sexual transmitted disease; it transmits also from mother to child during pregnancy, delivery, and breast feeding. Although its prevalence is low (less than 0.2%) in the Middle East and North Africa, the fear of stigma makes people reluctant to seek testing for this disease [15,16]. The disease-related stigmatization has impacts on HIV positive patients. They feel the humiliating and insulting looks by the people around who think that AIDS emerges from sexual deviances. HIV positive patients face isolation from family members, friends and relatives and experience discrimination in receiving health care. They feel separated, lonely, hopeless and rejected by the community [17]. The majority of HIV positive patients do not know they have the virus. Treatment of AIDS using antiretroviral therapy (ART) decreases greatly the risk of HIV transmission between partners and decreases dramatically the transmission from mother to her child as well [18,19].

Consanguineous marriages are costly

Genetic disorders, tend to be chronic, difficult and expensive to treat, and sometimes life-threatening. β -thalassemia for example costs more than USD 400,000 per person over a lifetime. Cystic fibrosis costs USD 9,400 per person per year; the overall cost of almost all common genetic disorders in the whole Arab World is estimated to be USD 13 billion per year. Many of these diseases are physically or mentally disabling, and some are terminal. These diseases impact the productivity of the future workforce and requires the government allocate more resources-for example special education services. Some of these diseases have very harmful effects on the emotional wellbeing of the family. Patients afflicted with cystic fibrosis should be prepared to die by around age 30 or perhaps earlier [20]. Mackenzie et al. [21] reported that median survival of cystic fibrosis patient in the United States is 37 for males and 40 for females.

Premarital screening from a human right's perspective

The consanguineous marriages are so common and favored in the Middle East for many reasons. These marriages may

strengthen the couples' stability thanks to the higher compatibility between the spouses who share the same social relationships after marriage as before marriage and the compatibility between the spouses and their family members as well [4].

Consanguineous marriages might be more favorable for women, thanks to the wife's better relationship with her in-laws who could support her in the hard times. There is a general belief that consanguineous marriages reduce the likelihood of hidden uncertainties in financial and health issues, and that consanguinity fosters family ties and strengthens family solidarity [22]. Premarital negotiations regarding financial issues of marriage are more easily carried out. Wife's parents prefer to see their daughter residing near them and enjoy the presence of their grandchildren as well. Moreover, rich landlords may prefer to keep their properties within the family [4].

Islam (which is the main religion in the Middle East) does not favor unions between close relatives; on the contrary, Islam discourages marriages to cousins who, because of the closeness of the relationship, are almost like siblings. In fact, the custom of consanguinity has nothing to do with Islam as many believe; it is just a tradition [23].

Given the impacts of consanguineous marriages on public health, it is important to provide health education on consanguinity at individual, family and community levels delivered mainly by the state's primary health care staff. It is also important to provide pre-marriage and pre-pregnancy genetic diagnosis and counseling (in case either or both of partners/spouses carries a genetic disease). Consanguineous marriage is a deep-rooted and long-held tradition in the Middle East. And because it is hard to discourage this type of marriages, ensuring access to pre-pregnancy and pre-marriage counseling services would be the logical way to go with. These services would be accepted by the community and would be successful in maintaining and enhancing health [24]. Increasing public literacy on consanguineous marriages could be done by providing appropriate education and training to primary health care personnel on all health and social issues related to relative marriages [4].

3.10) Premarital screening and the right to health

According to the International Covenant on Economic, Social and Cultural Rights (ICESCR)- adopted by the General Assembly of the United Nations on 16 December 1966 and monitored by the United Nations' Committee on Economic, Social and Cultural Rights – the Right to Health is “the right of everyone to the enjoyment of the highest attainable standard of physical and mental health” taking into consideration the individual's preconditions (biological and socioeconomic) and the country's available resources. The right to prevention, treatment and control of diseases (ICESCR, Article 12.2 (c)) “requires the establishment of prevention and education programs for behavior-related health concerns such as sexually transmitted diseases, in particular HIV/AIDS, and those adversely affecting sexual and reproductive health, and the promotion of social determinants of good health” [25].

The United Nations' Convention on the Rights of the Child (adopted in 1989) states that “the family, as the fundamental group of society and the natural environment for the growth and well-being of all its members and particularly children, should be afforded the necessary protection and assistance so that it can fully assume its responsibilities within the community” [25].

The Availability, Accessibility, Acceptability and Quality (AAAQ) framework should be sufficient to implement the Right to Health in this regard (WHO, The Right to Health). Availability means specialized facilities/centers for pre-marriage and pre-pregnancy genetic counseling and testing services should be sufficiently available within the country. Accessibility means these facilities should be within safe physical reach, financially affordable- premarital screening is costly; governments should provide it for free of charge, or at discount price- for all consanguineous couples who plan to get married soon, and should be accessible without discrimination. Information on consanguinity (and its impacts on public health) and services provided for consanguineous couples who are planning to get married soon should be accessible (through television, radio, internet, newspapers, mobile applications, posters, etc.). Accessibility may vary between rural and urban areas; between impoverished and prosperous neighborhoods, among the rich and poor. Acceptability means the abovementioned services should respect medical ethics and be culturally appropriate, i.e. sensitive to gender requirements and confidentiality of premarital screening results and personal health data. Quality means the abovementioned services should be scientifically and medically appropriate, and of good quality; this requires trained health personnel and diagnostic equipment; the quality is measured from the perspectives of health system as well as the consanguineous couples.

To marry and produce is a fundamental human right. Article 23 of the International Covenant on Civil and Political Rights states that “The right of men and women of marriageable age to marry and to found a family shall be recognized” (OHCHR, International Covenant on Civil and Political Rights) [26]. Building on this, human rights principles shall guide the whole aspects of premarital screening program to include planning, design (i.e. setting of strategies, goals and objectives), execution, monitoring and evaluation. Among those principles are non-discrimination, participation, inclusion, and accountability. (UNICEF, Introduction to the Human Rights based Approach). Premarital screening program personnel may take introductory course in this human rights-based approach as well.

All parties should be involved in this program (WHO, A Human Rights-based Approach to Health)- politicians (to enact laws), religious leaders/clerics (to legalize/reconsider therapeutic abortion in case the fetus afflicted/diagnosed with grave congenital malformation), employees in primary health care, researchers, doctors, geneticists, disease prevention and control and social services, subject-matter experts, mother and child health care, media, and activists in NGOs and voluntary organizations.

Monitoring and evaluation of the program should be expressed in terms of progress being made in achieving outcomes with regard to allocated budget and government spending on the program, percentage of consanguineous couples who have access to the screening program, cost-effectiveness analysis, rates of infectious diseases (e.g. hepatitis B, hepatitis C, and HIV/AIDS), genetic-recessive diseases (e.g. thalassemia, sickle cell anemia and cystic fibrosis), congenital malformations among children, inborn errors of metabolism diseases (e.g. mucopolysaccharidosis, phenylketonuria), multifactorial disorders (mental retardation, asthma, diabetes and epilepsy) and sensorineural deafness, consanguineous couples' satisfaction, etc [27].

Ahmadnezhad et al. [28] reported from Iran that premarital and prenatal screening for thalassemia proved to be cost-effective. They found that detecting high-risk couples or affected embryos and terminating pregnancies cost less than treating thalassemia. They recommended such screening programs are provided in the public and private sectors.

Conclusion

Consanguineous marriages are so common in the Middle East and North Africa although Islam has nothing to do with this long-held deep-rooted tradition. Being a part of the culture of Middle Easterners, it is not easy to change their views on this type of marriages. Raising awareness about the impacts of consanguineous marriages on public health is time-consuming process. However, it is important to address this issue through premarital screening which aims to advise couples (first-cousins in particular) who are planning to get married soon on the odds of transmitting a group of diseases to the other spouse, or to their children, and to provide couples with options that help them plan for healthy family. Premarital screening should be guided by human rights principles to include non-discrimination, participation, inclusion, and accountability. Moreover, the Availability, Accessibility, Acceptability and Quality (AAAQ) framework should be sufficient to implement the Right to Health in this regard.

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