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Research Article

Safe Marriage for Thalassemia Prevention; the Gap between Knowledge and Practice among Medical Students

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Abstract

Thalassemia is the commonest recessively inherited chronic haemolytic anaemia. Sri Lanka records 1500-2000 thalassemia patients, 1-2% carriers and 80 new cases per year. The government sponsored lifelong palliative treatment accumulates cases escalating the cost of care that mandates prevention. Law prohibits antenatal diagnosis and abortion of affected babies. However one of the partners in a couple been a non-thalassemia carrier eliminates the risk of having affected babies. Such marriages are called 'safe marriage' in Sri Lankan thalassemia prevention programme. Screening and decision making is voluntary and impetus is entirely dependent on public education. How much could be achieved by education alone has not been tested. Medical students are the most educated clients of this programme. Their practices were expected to indicate the success achievable in this prevention strategy by education alone.

Method: A pretested questioner was administered to first and final year medical students after ethical approval.

Results: Out of 89.9% first and 83.5% final year students did not know about their thalassemia status. Among those with affairs, 89.5% of first years and 83.3% of final years did not know their thalassemia status. However, 87.4% students' recommended compulsory screening and 73.7% recommend ending affairs at-risk, 45.5% indicated that such advices are inhuman and 16.4% recommend proceeding with marriage but avoiding baring children. Regarding monitoring marriages at risk 81.1% recommended the Registrar of Marriages to inquire about it at the time of marriage and 72.3% recommended referral of those couples for medical advice. Only 39.4% promoted legalisation of abortions, but 61.8% statements rationalized abortions and only 19.8% stated that it is inhuman or unjust.

Conclusion: In spite of having adequate knowledge, medical students do not practice the concepts of safe marriage. Therefore imparting knowledge alone may not be sufficient to achieve targets of the thalassemia prevention by the promoting safe marriage. However medical students endorse compulsory screening, directive counselling and abortions. Compulsory screening is mandatory for thalassemia prevention in Sri Lanka.

Keywords: Thalassemia Prevention; Safe Marriages; Knowledge Practice Gap

Introduction

Earliest reports of association between abnormally shaped red cells and anaemia in the early nineties; by Herrick in 1910, Huck in 1923, Dreshback in 1904, Cooley and Lee in 1926 were not termed thalassemia [1]. Severe anaemia among children of Italian origin was named Cooley's anaemia after Thomas Cooley and Pearl Lee who reported these cases [2,3]. Later this condition was termed thalassemia; the Greek word 'thalasa ' denotes sea, as the disease mainly affected children of Mediterranean origin; around the sea. However the disease is no more confined to Mediterranean region due to improvements in international travel and migration. Thalassemia is the commonest genetic disorder worldwide. Of the world population 4.83 % carry defective globin genes, resulting in symptomatic disorders in 2.4 per 1000 births, out of which 0.44% have thalassemia [4]. Thalassemia's include a wide spectrum of diseases. This discussion is confined to beta thalassemia, which constitutes a major problem in Sri Lanka.

In thalassemia the premature destruction of red cells that carry haemoglobin occurs and this results in anaemia. In a normal person, an enormous number of red cells are produced constantly from the bone marrow to maintain 500 million red cells per ml in 5 liters of blood in an adult human body. The entire haemoglobin content in our body is found within the red cells and contribute for the redness that is reflected by normal haemoglobin level of 14-16 gr per deciliter. In normal individuals, red cells survive up to 120 days but thalassemia red cells break down prematurely due a defect or a deficiency of one of the two types of protein chains found in the red cell. These protein

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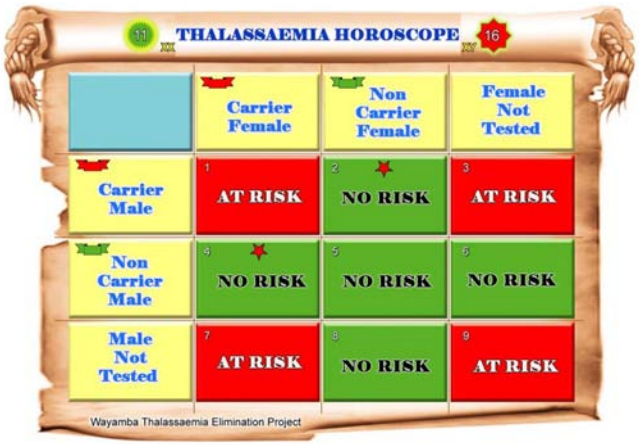

Figure 1: Thalassemia horoscope; an aid to decide whether a proposed couple is safe or at risk with regards to risk of having offspring with thalassemia.
Box number 2 and 4 is highlighted to indicate that those who select a known thalassemia carrier as his/her partner for the marriage should check their blood by HPLC to confirm that they are not thalassemia traits.

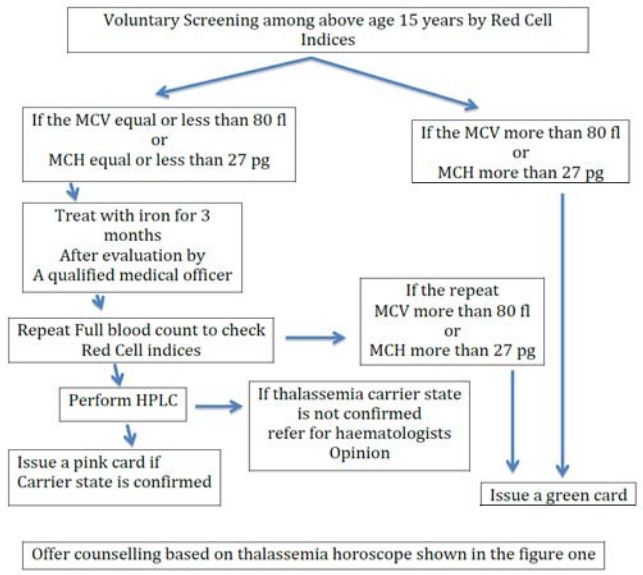

Figure 2: National Thalassemia Screening Protocol of Sri Lanka.

chains are called alpha or beta and thalassemia that results due to these deficiencies are called alpha and beta thalassemia.

The genes responsible for the production of globin component of the haemoglobin these proteins are located in the 11th or 16th pair of chromosomes respectively. When both genes in a pair of chromosome carry defects, disease status of thalassemia it is called thalassemia major and becomes discernible. These genetic defects are categorized as recessive disorders as those who have defective genes in one chromosome do not have significant implications of carrying this gene except for the possibility of passing these defective genes to the next generation; therefore they are called thalassemia traits or carriers. Marriages between two thalassemia carriers carry the risk of having offspring with major manifestation of thalassemia. As such avoiding marriage between two carriers is the basis of thalassemia prevention within the concept of safe marriage.

In Sri Lanka the carrier rate varies from 1% up to 5% [5]. An estimated national annual incidence 80-100 cases with 20-25 years of average survival results in a disease burden of 1600-2000 cases in the country. Base on these numbers on the assumption of 2 children per couple, it is predicted that there would be 160–200 marriages at risk every year in the country. All the marriages are registered in the country and about 150000 marriages take place every year. The incidence of marriages at risk would be around 1.6 - 2 per one thousand marriages.

Patients with thalassemia major require regular lifelong blood transfusion for survival, which results in inadvertent accumulation of iron in the recipients as major portion of body iron is found in red cells. Iron is toxic and there is no natural mechanism of iron removal from the body. Therefore all those who receive regular blood transfusions require regular lifelong medication to remove iron from the body, ultimately costing 250000 – 1000000 USD per patient in the UK [6] and about 2465 USD per patient per year in Sri Lanka [5]. Improved efficacy of care has resulted in improved survival and increased case prevalence. The cost of care escalates due to the increased number of cases as well as the higher cost of care for bigger children making thalassemia prevention imperative.

The concept of safe marriage recognizes a proposed couple as 'safe' if one person in the couple is not a carrier and portrayed in a figure simulating a horoscope (Figure 1). Sociocultural and economic implications of this concept have been evaluated [7]. And screening protocol (Figure 2) and facilities have been established before the national thalassemia prevention program was launched in 2008. The National Thalassemia Committee recognizes four indicators for monitoring and guiding the thalassemia prevention program. (Table 1). Motivation of the public to adhere to the concept of safe marriage is a challenge. The National Thalassemia Committee embarked on expansion of knowledge in public and schools in the country. However how much could be achieved by education alone is a concern. This study evaluation of the behaviour of medical students as a group of people with comprehensive knowledge on thalassemia with regard to safe marriage concepts was considered as an indicators of success that could be achieved by education alone.

Methodology

A questionnaire was developed by consensus of a group of experts to evaluate the practices and attitudes of medical students regarding safe marriage for prevention of thalassemia. The initial questionnaire was pretested for its comprehension and validity with a small group of students. A pretested questionnaire was administered to the first and final year medical students after obtaining ethical approval and the permission from the dean at the end of a routine lecture. Students were given the option to respond or not. Senior investigator was not present at the time of administration of the questionnaire promoting the free expression of their opinion. The responses were analysed and the proportions were calculated.

Results

Descriptive analysis of the survey data is presented in (Table 1). The opinion of the respondents relevant to thalassemia prevention

Table 1: Monitoring indicators of the national thalassemia prevention project of Sri Lanka recommended by the National Thalassemia Committee.

	How to monitor	Remedial Action
1. Percentage of screening coverage	To be monitored by Public Health Midwives	Education promotion and providing facilities for screening
2. Incidence of at risk marriages	To be monitored by registrar of marriages	Providing feedback for respective health care teams at district level
3. Incidence of at risk pregnancies	To be monitored by the Obstetrician at the antenatal clinics	Providing feedback to respective health teams at district level
4. Incidence of births of thalassemia major patients	By establishing a national register of thalassemia	Provide feedback at regional and national level

Table 2: Descriptive summary of the respondents according to first and final year medical students of the Faculty of Medicine Peradeniya in 2013.

	1st year students	Final year student
Total number responded (response rate)	189 (95%)	97 (48%)
Male	80 (42.3%)	42 (43.29%)
Female	109 (57.6%)	55 (56.7%)
Having relatives with thalassemia	11 (5.8%)	3(1.0%
Having seen a patient with thalassemia	60 (31.74)	97 (100%)
Having an affair	52 (30.2%)	51 (52.7%)
Their affair is planned to end up in a marriage	42 (73.6%)	51(92.1%)
Have you tested your blood? –Response was NO	170 (89.9%)	81(83.5%)
Having good knowledge about thalassemia	122(64.4%)	97 (100%)
Total number of siblings of respondents	265	140
Total number of siblings tested for thalassemia	11(4.15%)	15(10.7%)

is presented in (Tables 2-8). Among 30.2 % first year and 52.7% final year students who have selected their partners, only 10.5% and 18.7% had tested their blood for thalassemia (Table 1), whilst 72.8% of the above group recommend ending affairs at risk (Table 1 and 4). When we consider 89. 4% of the first years and 86% final years who recommend some form of compulsory screening only 10.2 % has screened themselves (Table 1 and 2), and only 4.6% had screened their own siblings (Table 1 and 2).

Discussion

Thalassemia is a major health care problem in Sri Lanka. The problem of health care delivery for those who are suffering from thalassemia is complex. Affected children and their families suffer their entire life as the only curative therapy of bone marrow transplant is not affordable to the majority. The escalating cost of sponsored health care burden on the government is further escalates due to the nature of palliative treatment offered. Thalassemia prevention strategies are confined to teenage counselling, as other widely practiced methods are unlikely to become established in the country in the near future. Prenatal diagnosis and abortions has been established as the most effective method of thalassemia prevention in many countries [8], however this approach is not possible due to legal

Table 3: Opinion of the respondents regarding thalassemia screening.

	1st year students	Final year students
Thalassemia screening should be made compulsory	107 (56.6%)	42 (43.3%)
Only voluntary screening should be promoted	17 (8.99%)	14 (14.4%)
Screening of at least one of the partners in a couple should be made compulsory	22 (11.64%)	25 (25.8%)
Both partners in a couple should be screened	43(22.8%)	16 (16.5%)

Table 4: Opinion of the respondents regarding the age of screening for thalassemia.

	1st year students	Final year students
During the school days	104(55.0%)	67 (69.0%)
After school but before a marriage proposal	56(29.6%)	21(21.6%)
After proposal but before the marriage	27 (14.3%)	8 (8.2%)
After marriage but before the pregnancy	2 (1%)	1 (1%)

Table 5: Opinion regarding advice to thalassemia carriers, non-carriers and couples when both are carriers (Questions 16, 17 and 18).

Q16- What is your advice for a thalassemia carrier	1st year students	Final year students
16.1 do not embark on a marriage	3 (1.5%)	0(-)
16.2 can marry with a non carrier	149 (78.8%)	87(89.7%)
16.3 can marry but avoid having children	37 (19.6%)	10 (10.3%)
Q17- What is your advice for a person who is NOT a thalassemia carrier		
17.1 do not marry a carrier	23 (12.2%)	3 (3.1%)
17.2 marry only with a non carrier	37 (19.6%)	12 (12.4%)
17.3 can marry anyone	129 (68.3%)	82 (84.5%)
Q18-What is your opinion when both partners of a couple are thalassemia carriers		
18.1 We should let them decide what to do	8 (4.2%%	12 (12.4%)
18.2 Counsel and advice to avoid that marriage	143 (75.7%)	68 (70.1%)
18.3 Advice antenatal diagnosis and abortions	33 (17.5%)	15 (15.5%)
18.4 Marriage between carriers should be sanctioned by law	5(2.6%)	2(2.1%)

sanctions on abortions which is unlikely to get rectified due to the influence of religions, socio cultural background. Facilities for other advanced methods of thalassemia prevention like pre implantation diagnosis and in vitro fertilization are not yet available.

The concept of safe marriage has its own strengths. The approach is scientific and message is simple and integrates with the established concept of horoscope reading in the societies of the region. Horoscope reading is utilized to assess the suitability of a partner. The entire process empowers the society allowing them to make informed decisions on the most important events in a person's life. There are no punitive actions or compulsions incorporated into the program. However consequences of non-adherence will act as a natural reinforcement. The health care professionals role would be to offer support and guide the society in correct directions. The process of screening has a sound scientific basis.

Screening and surveillance of disease conditions in a community is appropriate when the condition has known natural history, a latent phase, suitable and acceptable test and an effective remedy [9]. Screening program should have an agreed policy, facilities

Table 6: Opinion regarding the proposed monitoring process by the National Thalassemia Committee at the time of the registration of marriage by the registrar.

Q19- Inquiring about the carrier state of the couple by a registrar of marriage at the registration	1st year students	Final year students
19.1 Should not be done	29 (15.3%)	24 (24.7%)
19.2 Inquiry should be done confidentially and individually	69 (36.5%)	19 (19.6%)
19.3 Inquiry should be done openly	85 (45.0%)	47 (48.5%)
19.4 If the male is a carrier inquire the female	6 (3.1%)	7 (7.2%)
Q20- what should be done by the registrar regarding an at risk marriage		
20.1 do not register	5 (1.6%)	3 (3.1%)
20.2 refer to a medical officer and reconsider	143 (75.7%)	64 (66.0%)
20.3 register them without intervening	5 (2.6%)	7 (7.2%)
20.4 allow them to reconsider after counseling	36 (19%)	23 (23.7%)

Table 7: Opinion regarding counseling of at risk couples.

Q21- Your opinion on directive counseling an at risk couple to avoid that marriage	1st year students	Final year students
21.1 it is not justifiable	29 (13.2%)	28 (18.4%)
21.2 It is a violation of basic human rights	16 (7.2%)	18 (11.8%)
21.3 it is inhuman	18 (8.1%)	21 (13.8%)
21.4 it is a commitment for the benefit of future generation	146 (66.4%)	66 (43.4%)
21.5 it is a commitment of a thalassemia carrier towards the society	11 (5%)	19 (12.5%)

Table 8: Opinion regarding abortions for prevention of thalassemia.

Q22- What is your opinion regarding abortion of a foetus with thalassemia	1st year students	Final year students
22.1 it is inhuman	37 (10.7%)	21 (11.4%)
22.2 its an injustice for the foetus	35 (10.1%)	12 (6.4%)
22.3 its a justice for the foetus	74 (21.4%)	29 (15.7%)
22.4 it is justice for the mother and the family	70 (20.3%)	47 (25.4%)
22.5 it is beneficial to the country	73 (21.2%)	38 (20.5%)
22.6 restrictions for abortions in such occasions should be lifted	56 (16.2%)	38 (20.5%)

for screening and an established on-going cost effective process as explained in Wilsons and Jungner principals. Thalassemia carrier status is a condition that fulfils all these criteria. Thalassemia carriers are having a recognizable latent condition with a risk of having offspring suffering from a major disease with a well-known natural history. The test available for screening is cost effective and acceptable for the society. Once the problem is identified practical solutions are available; either to avoid an at risk marriage, avoid conceptions or explore possibilities of antenatal diagnosis. However the last option that should be coupled with abortion of affected babies is not practiced in the country due to the barriers mentioned earlier. There is an agreed policy for screening, however the method of management is confined to the avoidance of marriages at risk. The cost of case detection seems to be reasonable and preliminary screening by the simple blood count is possible all over the country and confirmation tests offered to about 20% of those who come for screening are given the services of the screening centers in the 5 general hospitals and this has ensured the continuity of screening by the government of Sri Lanka. However integrating the process into the system seems slow. The monitoring indicators proposed by the NTC have not been fully implemented. Therefore the progress of the project has not created the expected impact on society as yet. The question whether education alone is sufficient for the expected paradigm shift in the society remains unanswered.

Medical students form a group in the society with the best of knowledge on thalassemia and methods of prevention. Evaluation of their behaviour creates an understanding on some aspects of human behavior regarding risk taking related to health. The gap between knowing and doing is a well-known phenomenon in human behavior. Knowing the risk of a particular behaviour does not necessarily result in changing that behavior. In this example, medical student's knowledge seems to have motivated them to recommend compulsory screening but they have not tested their blood or their siblings. Those who have initiated affair with the intention of marriage have not tested their blood while a majority of them recommend ending affairs at risk. Similar behaviour with regard to health care advice is observed in many community based prevention programs (Eden 2002) such as the prevention of accidents obesity, alcohol smoking and cancer, birth defects and thalassemia prevention [10-15].

Deeper analysis of the reasons for failure in adherence of the public should be analyzed based on systematic-heuristic theoretical model of decision-making. Heuristic factors are very important in decision-making.

Mandatory screening and counselling as endorsed by majority of our students in this study and in similar opinion surveys in the past [7]. Mandatory screening has helped many countries to succeed in implementing thalassemia prevention programs such as Cyprus Iran China Israel and Jedah [16-20].

Non-directive counseling is advocates to ensure the right and ownership of decision making to the counselee [21]. Motivational interviewing is an acceptable method of convincing people on avoiding health risks [22]. Majority of our students recommends counseling to avoid at risk marriages as well as terminate affected babes. Whether these attitudes constitute directive counseling is arguable. Non-directedness should not preclude divulging the entire natural history of the condition. Therefore the process of divulging information on the risks of thalassemia carriers' should be acceptable even though it may sound likes directive counselling. Counselors attempt to alleviate their clients by depicting thalassemia as a condition that is not so difficult to manage is deceiving and far from the real truth at least in the developing countries. Therefore counselling of thalassemia carriers will turn out to be directive counselling due to the nature of the disease in the context of our society.

Sri Lanka has been successful in community based prevention programs due its high literacy ratio and the well-established primary health care system. The Vaccination program is very successful and exceeds coverage over 90% even in the regions affected by war during the war [23]. Acceptance of vaccines did not dent even after several incidences of adverse effects and two deaths following the vaccine [23]. Therefore the findings of this study do not hamper the possibilities of implementing community based thalassemia prevention program but emphasises the need for a comprehensive and integrated approach. Any community based surveillance and prevention program needs monitoring.

Conclusion

Medical students do not practice the concepts of safe marriage in spite of having adequate knowledge. Therefore education alone is not sufficient to change the behaviour of population with regard to the safe marriage concept. However they endorse compulsory screening, directive counselling and abortion. These finding needs further clarification with a representative sample in the society and a wider discussion to eliminate the bias of the perceptions of a medically oriented group.

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Ethical Statement

Ethical clearance was obtained from the ethical review committee of the Faculty of Medicine Peradeniya.

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