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Original Article

Acceptance towards giving birth to a child with beta-thalassemia major – A prospective study

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ABSTRACT

Objective: To explore the acceptance of pregnant Chinese women on giving birth to a child with beta-thalassemia major.

Materials and methods: Women's acceptance on having a child with beta thalassemia major was assessed using standard gamble metrics during an interviewer-administered survey on 309 women recruited in the antenatal clinic. Utility scores were determined and the association with sociodemographic factors was assessed.

Results: The median utility score for having a child with beta-thalassemia major was 0.5 (0–0.65). Women having either higher educational level or family income had significantly higher utility scores ($P < 0.05$) corresponding to a higher acceptance. During the interview 59.9% participants indicated that they would elect to undergo a termination of pregnancy if their fetus was diagnosed with beta-thalassemia major but 26.5% participants were unable to decide what action they would take.

Conclusion: Many Chinese pregnant women are ambivalent about giving birth to a baby with beta-thalassemia major. Women with higher educational level or higher family income had a higher acceptance towards the condition.

Introduction

Beta-thalassemia is a common inherited disorder of hemoglobin synthesis with a reported carrier frequency in Southeast and East Asia ranging from 2 to 9% [1]. Beta-thalassemia is caused by a mutation in the HBB gene located on the short arm of chromosome 11 which controls production of the beta-globulin protein. The loss or impaired production of beta-globulin results in red blood cell damage and anemia. In the homozygous state, marked reduction of functional hemoglobin levels results in transfusion dependent anemia, reduced growth, delayed development, bone abnormalities and hepatosplenomegaly. The mainstay of treatment currently is lifelong transfusion with concomitant iron chelation therapy to prevent the adverse effect of transfusion-mediated iron overloading and hemochromatosis, while successful allogeneic hematopoietic stem cell transplantation has been demonstrated in selected cases [2,3]. Mortality rates are reduced from 12.7 to 4.3 deaths per 1000 patient as therapies improved and life span has increased from 20 to 45 years since the 1970s [4].

Beta-thalassemia is an autosomal recessive inherited condition. Offspring of beta-thalassemia couple have a 25% chance of inheriting both mutations from the parents and being affected with beta-thalassemia major. Antenatal screening using mean corpuscular volume is effective in identifying asymptomatic carriers and at risk pregnancies [5]. Ultrasound screening however is unable to detect beta-thalassemia major prenatally because features of the disease only occur after birth unlike in the case of alpha-thalassemia major. Fetal diagnosis of beta-thalassemia major is reliant on genetic studies on fetal tissue obtained through either chorionic villus sampling or amniocentesis. Both of these diagnostic procedures are associated with procedure related fetal loss, with loss rates commonly quoted as ranging from 0.5 to 1% during counseling, although more recent studies have indicated rates as low as 0.1% [6–9].

The attitude towards giving birth to a child with abnormalities varies in different cultures. Chinese women who often find a handicapped child unacceptable have a low tolerance of fetal

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chromosomal abnormalities as demonstrated by the high proportion opting for elective termination of pregnancy for fetus diagnosed with Down syndrome [10,11]. In contrast to Down syndrome, our empirical observation would suggest that some Chinese women opted to continue pregnancies diagnosed with fetal beta-thalassemia major prenatally after counseling by Pediatric Hematologist on the prognosis and management options. We postulate that this may be because beta-thalassemia major is not associated with impaired intelligence unlike Down syndrome. There are no published studies on the acceptance of Chinese women's towards giving birth to a child with beta-thalassemia major. Since beta thalassemia is a relatively common inherited condition in our locality, understanding to what extent prospective parents accept, and the factors that could influence their choice, could help with effective prenatal counseling.

The method of Standard gamble metrics was previously used to assess the acceptance of Chinese women on having a Down syndrome baby and the metrics was well understood by the participants [11]. The study was able to demonstrate that Chinese women perceived having a Down syndrome baby a similar adverse life event as being blind and they had a much lower acceptance towards Down syndrome baby compared to Caucasian women [11]. Thus, standard gamble metrics was used in this study to assess Chinese women's acceptance on having a baby with beta-thalassemia major.

Materials and methods

This was a prospective observational study conducted in a University unit in Hong Kong which offers universal first trimester combined screening test for Down syndrome [12]. Women who attended the clinic for assessment of fetal Down syndrome risk over a 4 month period were invited to participate in an interviewer-administered survey. All interviews were performed by a single individual with previous experience on administering the standard gamble technique [11]. Women who were non-Chinese or who did not speak Chinese were excluded from the study. The study was approved by the Institutional Ethics Committee and informed consent was taken from all subjects.

Before the survey was conducted, women were provided with written information on beta-thalassemia major including its genetic basis, cause, presentation and potential management options including regular blood transfusion, iron chelation therapy, and hemopoietic stem cell transplantation as well as current option to diagnose whether their fetus was affected. Women were informed that beta-thalassemia major could be diagnosed by an invasive diagnostic procedure with an associated risk of miscarriage of around 0.5–1%.

The survey was composed of two sections. The first section was designed to elicit relevant demographic information, including maternal age, marital status, educational history, family income level, obstetric history and family history. The women's attitude towards fetuses with beta thalassemia major and termination of pregnancy was assessed by the following question. 'Would you request a termination of pregnancy if your fetus is diagnosed with beta thalassemia major.'

The second section was designed to elicit patient acceptances using the standard gamble metrics. In a standard gamble test, subjects will be asked to choose from a guaranteed outcome (e.g. being blind for the rest of their life), or whether they would prefer an alternative state with some chance of a worst outcome (e.g. dying, '1-p') and a complementary chance of an ideal outcome (e.g. continue to live healthily, 'p'). The risks for the two situations are varied until the subject is indifferent to either of the two options, at which stage the 'utility' for the health state under consideration equals 'p'. This allows quantitative comparison of qualitatively different health states, which guides our understanding of the decision making process of the patients.

In order to ensure that our subjects understood the standard gamble metrics, the interviewer first assessed their utilities towards the health state of blindness. The subject was asked to make a decision starting with a probability of 100%, which was then reduced in steps of 5% until it reached 10% after which it was decreased at 1% interval until 2% and thereafter at 0.1% interval, until the subject was unable to make a choice between the two options. After ensuring that subjects understood the standard gamble methodology, the exercise was repeated and the utility towards pregnancy affected by Beta-thalassemia major is assessed using the same approach. The lower the utility, the lower was the patient's acceptance towards that health state.

Utility scores were summarized by median and interquartile range (IQR). Difference in scores between different patient groups was compared using Mann–Whitney U test. The Statistical Package for Social Sciences for Windows version 20 (IBM, Armonk, NY, USA) was used for statistical analysis of all data. A p-value of <0.05 was considered statistically significant.

Results

A total of 315 women who attended the clinic for first-trimester Down syndrome screening were invited to participate in the study, all agreed. Six women subsequently refused to complete the survey as they could not comprehend how the standard gamble assessment was performed, and were unable to complete the blindness standard gamble pre-assessment. These women were excluded from all further analyses. The 309 remaining participants were able to understand the standard gamble metrics and completed the survey.

The demographic characteristics of these 309 women are summarized in Table 1. Nine women were known to be alpha- ($n = 7$) or beta- ($n = 2$) thalassemia carriers prior to conducting the survey. A further 11 women were identified as alpha- ($n = 7$) or beta- ($n = 4$) thalassemia carrier after completing the survey by antenatal screening which indicated decreased MCV, and which was subsequently confirmed by hemoglobin pattern testing. The MCV of their partners were normal except that for the partner of one alpha-thalassemia carrier who was found also to have low MCV and was subsequently confirmed as being a beta-thalassemia minor carrier by hemoglobin pattern assessment.

Table 1
Demographic characteristics of the study population. Values are expressed as mean ± SD or as percentage.

Characteristic	
Age, years	31.9 ± 4.5
Married	92.6%
Education	
Secondary or below	55%
Tertiary or above	45%
Family income (HKD per month)[a]	
<50,000	80.6%
≥50,000	19.4%
Previous miscarriage	17.8%
Previous delivery	42.7%
Previous pregnancy termination	24.6%
Previous neonatal death	0
Previous stillbirth	0
Relative or friends with beta thalassemia	1.6%
Known thalassemia carrier	3.6%
IVF pregnancy	3.2%

[a] HKD 1 = USD 0.1281.