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REVIEW ARTICLE: THALASSEMIA IN ASIA 2021

Thalassemia Status in Cambodia

Chean Sophâl

Department of Pediatric Hemato-Immunology, National Pediatric Hospital, Phnom Penh, Cambodia

ABSTRACT

The population of Cambodia (in 2019) was approximately 16 million with an annual growth rate of 1.4% in which the prevalence of hemoglobinopathies was estimated at about 40.0% (range 30.0–50.0%) to be carriers, and 2240 annual births for β-thalassemia major (β-TM). The overall prevalence of β-thalassemia (β-thal) and α-thalassemia (α-thal) were 40.9 and 39.6%, respectively. Currently, the specific epidemiological data regarding the abnormal gene frequency/mutations among different ethnic groups is unknown. In 2011, national guidelines for the Clinical Management of Patients with Thalassemia in Cambodia were developed and published by the Ministry of Health (MoH). Packed red cells (PRCs) are available at most referral hospitals (provincial hospitals). Oral iron chelators [deferiprone (DFP) and deferasirox (DFX)] are only available from a private pharmaceutical company. The future needs for Cambodia are to develop a national policy on the prevention or control of β-thal and α-thal, and a national registry of patients with thalassemia, to determine the gene frequency of α- and β-thal in different regions of the country, and to place the iron chelators on the list of essential medicines.

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Cambodia; prevalence; thalassemia

The population of Cambodia (in 2019) was approximately 16 million with an annual growth rate of 1.4% [1]. Life expectancy at birth is 75.5 years and under 5 years of age mortality rate is 28/1000 live births [1]. Approximately 96.0% of the Cambodian population is of Khmer ethnicity and speaks the Khmer language [1]. Gross national income per capita (in 2019) was US$4310.00 (Cambodian riel: 17,240,000.00) [2]; gross domestic product (GDP) per capita in 2019 was US$1643.12 (Cambodian riel: 6,572,480.00) [3].

With regard to the prevalence of hemoglobinopathies in Cambodia, about 40.0% (range 30.0–50.0%) of the general population were estimated to be carriers, of which 1.0% carry β-thalassemia (β-thal) [at least 4.8 million are carriers, 48,000 are β-thal major (β-TM) sufferers] [4–6]. The overall prevalence of β-thal and α-thalassemia (α-thal) were 40.9 and 39.6%, respectively [7]. The estimated annual birth rate of thalassemic patients is about 2240 births for β-TM. Currently, the specific epidemiological data regarding the abnormal gene frequency/mutations among different ethnic groups is unknown. A high prevalence of genetic hemoglobin (Hb) disorders exists in Cambodia (Table 1) [7].

In 2011, national guidelines for the Clinical Management of Patients with Thalassemia in Cambodia were developed and published by the Ministry of Health (MoH) [8]. In Cambodia, thalassemia is diagnosed by Hb electrophoresis using the Capillary Minicap Analyzer (Sebia, Lisses, France). Genotyping using molecular analysis is not available.

Transfusions

Packed red cells (PRCs) are available at most referral hospitals (provincial hospitals). The blood supply in the country is currently limited and most of the PRCs need to travel great distances to reach rural hospitals. Leukocyte-depleted packed red cells (LDPRC) are not available. Hydroxyurea (HU) is available at some private pharmacies. Blood group phenotyping and antibody screening are not available.

Chelation therapy

Oral deferiprone (DFP) and deferasirox (DFX) are available from a private pharmaceutical company, but are used on a very limited basis and is not presented on the list of essential medicine produced by the MoH. Notably, deferoxamine (DFO or Desferal®) is not available within the country.

Splenectomy

Splenectomy on severe thalassemia patients is carried out at some Phnom Penh and provincial hospitals. At least 3 weeks prior to splenectomy, the patients were immunized with pneumococcal, Hemophilus influenza B, and meningococcal vaccines. One clinical study done in 2012, at the National Pediatric Hospital, Phnom Penh, Cambodia, showed that 31.0% of severe thalassemia patients underwent the procedure, of which 11.0% carried β-TM and 20.0% carried α-thalassemia (α-thal) (Hb H disease) [9].

There is no national policy or program for the prevention or control of thalassemia in Cambodia apart from a general

CONTACT Dr. Chean Sophâl sophalces@yahoo.com Department of Pediatric Hemato-Immunology, National Pediatric Hospital, 100, Federation of Russia Boulevard, Phnom Penh, Cambodia

Table 1. Prevalence of different types of β- and α-thalassemia genotypes in Cambodia.

	HGVS Nomenclature	Prevalence (%)
β Genotype		
β^A/β^A	Normal	59.10
β^E/β^A	Heterozygous Hb E (*HBB*: c.79G>A)	34.52
β^E/β^E	Homozygous Hb E	6.07
$\beta^{IVS\text{-}II\text{-}654}/\beta^A$	IVS-II-654 (C>T) (*HBB*: c.316-197C>T)	0.06
$\beta^{IVS\text{-}I\text{-}1}/\beta^A$	IVS-I-1 (G>T) (*HBB*: c.92+1G>T)	0.12
β^{-28}/β^A	−28 (A>G) (*HBB*: c.-78A>G)	0.12
Total		100.00
α Genotype		
αα/αα	Normal	60.39
$--^{SEA}/\alpha\alpha$	Southeast Asian del	1.53
$-\alpha^{3.7}/\alpha\alpha$	3.7 kb rightward del	23.73
$-\alpha^{3.7}/-\alpha^{3.7}$	Homozygous 3.7 kb del	3.13
$-\alpha^{4.2}/\alpha\alpha$	4.2 kb leftward del	0.80
$-\alpha^{3.7}/-\alpha^{4.2}$	3.7 kb del + 4.2 kb del	0.12
$--^{SEA}/-\alpha^{3.7}$	SEA + 3.7 kb del	0.31
$\alpha^{CS}\alpha/\alpha\alpha$	Hb Constant Spring (Hb CS or *HBA2*: c.427T>C)	5.03
$\alpha^{Paksé}\alpha/\alpha\alpha$	Hb Paksé (*HBA2*: c.429A>T)	1.41
$-\alpha^{3.7}/\alpha^{CS}\alpha$	3.7 kb del + Hb CS	1.72
$-\alpha^{3.7}/\alpha^{Paksé}\alpha$	3.7 kb del + Hb Paksé	1.10
$-\alpha^{4.2}/\alpha^{CS}\alpha$	4.2 kb del + Hb CS	0.12
$-\alpha^{4.2}/\alpha^{Paksé}\alpha$	4.2 kb del + Hb Paksé	0.06
$\alpha^{CS}\alpha/\alpha^{CS}\alpha$	Homozygous Hb SC	0.31
$\alpha^{Paksé}\alpha/\alpha^{Paksé}\alpha$	Homozygous Hb Paksé	0.06
$\alpha^{CS}\alpha/\alpha^{Paksé}\alpha$	Hb CS + Hb Paksé	0.06
$--^{SEA}/\alpha^{CS}\alpha$	SEA del + Hb CS	0.06
$--^{SEA}/\alpha^{Paksé}\alpha$	SEA del + Hb Paksé	0.06
Total		100.00

commitment to reduce child mortality rates. A National Registry of patients, screening programs, and genetic counseling are not available in the country. Premarital screening is not routinely done. Due to technical problems, there is limited availability of prenatal diagnosis. In cases involving the termination of a pregnancy, there are no problems associated with legal, social and religious issues.

Future needs of Cambodia

The following are needed in Cambodia: (1) A national policy on the prevention or control of thalassemia, and a National Registry of patients with thalassemia, should be developed by the MoH. (2) New low-cost methods for detecting genetic Hb disorders are urgently required and the gene frequency of α- and β-thal in different regions of the country should be determined. (3) Iron chelators should be made available and provided by the MoH and placed on the list of essential medicines. (4) Support for the National Blood Transfusion Center to produce LDPRC, and (5) introduce procedures for blood group phenotyping and antibody screening.

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