# Progress Toward the Control and Management of the Thalassemias

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**KEYWORDS**

• Different forms of thalassemia • Screening • Prevention • Management
• National partnerships • Role of non-governmental organizations

**KEY POINTS**

- The prevention and management of the thalassemias is reasonably advanced in several countries in Asia.
- Partnerships are being developed between these countries and those in which facilities for the control and development of thalassemia are not yet established.
- There is urgent need for support on behalf of international health agencies for the further development of these programs.

## PROGRESS OF THALASSEMIA CONTROL IN THAILAND

### *Thalassemia in Thailand*

Both α-thalassemia and β-thalassemia and some abnormal hemoglobins (Hb), such as Hb E and Hb Constant Spring (CS), are prevalent in Thailand. The frequencies are 20% to 30% for α-thalassemia, 3% to 9% for β-thalassemia, 10% to 50% for Hb E, and 1% to 8% for Hb CS (**Table 1**).[1,2] Different combinations of these abnormal genes lead to more than 60 thalassemia syndromes. The 2 major alpha thalassemic diseases are Hb Bart's hydrops fetalis or homozygous $\alpha^0$-thalassemia and Hb H disease that results from the interaction between $\alpha^0$-thalassemia and $\alpha^+$-thalassemia or between $\alpha^0$-thalassemia and Hb CS. Almost all fetuses with Hb Bart's hydrops die intrautero or a few

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**Table 1**
**Thalassemia in Thailand**

| Major Types of Thalassemia | Frequencies, % |
|---|---|
| α-Thalassemia ($\alpha^0$-thalassemia and $\alpha^+$-thalassemia) | 20–30 |
| Hemoglobin Constant Spring ($\alpha^+$-thalassemialike effect) | 1–8 |
| β-thalassemia | 3–9 |
| Hemoglobin E | 10–53 |

minutes after birth and their mothers often suffer from obstetric complications, such as toxemia of pregnancy (preeclampsia and eclampsia), postpartum hemorrhage due to an enlarged placenta, and the psychological burden of carrying a nonviable fetus to term. Interaction between β-thalassemia genes or β-thalassemia and Hb E genes leads to homozygous β-thalassemia or β-thalassemia/Hb E disease, which are major beta thalassemic syndromes in this region. In β-thalassemia/Hb E disease, although the patients have identical genotypes, the degree of anemia varies greatly, with hemoglobin levels ranging from 3 to 13 g/dL.[3–5] The birth rates of major thalassemic diseases are shown in **Table 2**.

Since 2002, Thailand has implemented the Universal Health Care policy that resulted in a 99% universal coverage among Thai nationals using a mix of health protection schemes. Under this policy, all Thais will receive free treatment, either at the nearby district hospital or by referral for treatment in a medical center for cases that need special care. Almost all patients with thalassemia can have access to free blood transfusion and iron chelation, such as desferrioxamine and deferiprone. To reduce the cost of the iron chelators, the Government Pharmaceutical Organization has synthesized deferiprone for local use. The product is effective in the treatment of iron overload in all thalassemia syndromes, except major thalassemia cases that need regular blood transfusion that still need desferrioxamine treatment as a combination therapy.[6,7] The estimated direct cost for the management of a patient with major thalassemia who receives regular blood transfusion and desferrioxamine treatment who lives to be 10 to 30 years old is approximately 1.3 to 6.6 million Baht (US$ 39,393–200,000).[8] Even though thalassemia may be cured by stem cell transplantation, it is still expensive and it is difficult to find appropriate HLA-matched donors. Thus, the best approach to cope with thalassemia in

**Table 2**
**Total number of patients with thalassemia and the number of births per year (total births = 800,000/year)**

| Diseases | Couples at Risk (per year) | Births (per year) | Living Patients |
|---|---|---|---|
| Homozygous β-thalassemia | 828 | 207 | 2070 |
| β-Thalassemia/Hb E | 12,852 | 3213 | 96,390 |
| Hb Bart hydrops fetalis | 3332 | 833 | 0 |
| Hb H disease | 22,400 | 5600 | 336,000 |
| Total | 39,412 | 9853 | 434,460 |

*Abbreviation:* Hb, hemoglobin.
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