Hematological Practice in Hong Kong and China

Yok-Lam Kwong, MD[a], Shau-Yin Ha, FHKAM (Paediatrics)[b], Vivian Chan, PhD, FRCPath[a,*]

KEYWORDS

• Thalassemia • Hemophilia • Glucose-6-phosphate dehydrogenase deficiency
• Arsenic trioxide • Acute promyelocytic leukemia • Natural killer cell lymphoma

KEY POINTS

- Prenatal diagnoses of thalassemia, the most important inherited hematological disease in Hong Kong and Southern China, and hemophilia A and B have very significantly decreased the burden of these disorders.
- Adequate transfusion and iron chelation therapy for thalassemia major patients, and optimal factor replacement for hemophilia patients, have dramatically improved the outlook of these disorders.
- Arsenic trioxide is an active drug for acute promyelocytic leukemia on presentation and at relapse; the availability of an oral formulation means that long-term consolidation of remission with this drug can be achieved.
- Natural killer cell lymphomas, prevalent in Hong Kong and parts of China, have much better prognosis when treated with combination chemotherapy containing non-P-glycoprotein-dependent drugs and L-asparaginase.

INTRODUCTION

China has a population of 1.3 billion. Hong Kong is a special administrative region in China, with a population of 7.5 million. The Han ethnic group accounts for about 92% of the population in both places. Disease patterns in Hong Kong therefore closely reflect those in China.

The first major hematology center combining clinical and laboratory research was established in China in 1958.[1] In Hong Kong, research in blood diseases also started in the late 1950s. Allogeneic hematopoietic stem cell transplantation (HSCT) was first performed in 1981 in China,[2] and in 1990 in Hong Kong.

Disclosure Statement: The authors have nothing to disclose.
[a] Department of Medicine, Queen Mary Hospital, University of Hong Kong, Pokfulam Road, Hong Kong, China; [b] Department of Paediatrics and Adolescent Medicine, Queen Mary Hospital, Pokfulam Road, Hong Kong, China
* Corresponding author.
E-mail address: vnychana@hku.hk

Hematol Oncol Clin N Am 30 (2016) 445–456
http://dx.doi.org/10.1016/j.hoc.2015.11.010 hemonc.theclinics.com
0889-8588/16/$ – see front matter

Health Care Systems in China and Hong Kong

For a long time, patients or their employers were responsible for medical expenses in China. Recently, 3 systems of government-subsidized insurance schemes have been introduced, which cover up to 95% of the population. Hong Kong adopted a model similar to a national health care system, with patients paying a nominal daily fee of less than 15 US dollars even for complicated treatment such as HSCT.

However, with rising medical costs, such systems are becoming difficult to maintain. Whether the soaring fees should be met by increasing government expenditure or higher insurance premiums is hotly debated.

Burden of Hematological Diseases

Nonmalignant diseases including hemoglobin and bleeding disorders were predominant hematological problems in China and Hong Kong. However, malignant diseases now constitute the major burden, owing to an aging population and improvement in patient survivals.

NONMALIGNANT HEMATOLOGICAL DISEASES: GENETIC DISORDERS

The most common genetic diseases in this region are thalassemias (α- and β-thal) and glucose-6-phosphate dehydrogenase (G6PD) deficiency.

Thalassemias

The combined carrier rates for α^+- and α^0-thalassemia in Hong Kong are 4% and 3.5% for β-thalassemia minor.[3] In China, thalassemias are mainly restricted to the southern provinces of Guangxi and Guangdong, with α-thalassemia much more prevalent in Guangxi, with a carrier rate of up to 15%. In Guangdong, incidences of both types of thalassemias are similar to those in Hong Kong.[4] In Northern China, thalassemias are uncommon.

Prenatal Diagnosis for Thalassemias

Couples with the same thalassemia trait (α^0-thal or β-thal minor) carry a 25% risk of having a homozygous child. The homozygous α^0-thal fetus (Hb Barts hydrops fetalis) is incompatible with life. Early termination of pregnancy prevents maternal morbidity or even mortality. The homozygous β-thalassemia child (β-thalassemia major) lives, but is transfusion-dependent and suffers from the consequences of iron overload. Screening of at-risk couples in early pregnancy and subsequent prenatal diagnosis (PND) are advocated. Since 2000, screening is offered to all pregnant women on antenatal booking at every public hospital and maternity center in Hong Kong. A maternal mean corpuscular volume of less than 80 fl and a normal serum iron level necessitates investigation of the couple. In Hong Kong, PND was first performed in 1975 by globin-chain analysis of fetal blood. With the establishment of a DNA-based PND program in 1982, the detection of α-globin genes in fetal DNA excludes homozygous α^0-thal, but misses the occurrence of nondeletion Hb-H hydrops fetalis.[5] The various common nondeletion α-globin gene defects can be detected by reverse dot-blot or by microarray based on an allele-specific arrayed primer-extension technology[6]; this is important because nondeletion Hb-H accounts for 22.8% of Hb-H disease,[7] with at least 4 types giving rise to hydrops fetalis. However, α-Quong Sze (QS) and α-Constant Spring (CS), highly prevalent in Guangxi province and Southeast Asia, respectively, rarely cause hydrops fetalis. Most Hb-H patients are not transfusion-dependent and have a long lifespan.[7] Their only morbidity is increasing iron overload with age, which may then benefit from short-term iron chelation therapy.[8] Hence, termination of pregnancy is not justified for nonhydropic Hb-H fetus.