INTRODUCTION

Blood is a type of connective tissue that is an important part of the human circulatory system. It consists of red blood cells (RBCs), white blood cells (WBCs), platelets and several proteins, suspended in a yellowish liquid called plasma. Each RBC may contain between 240 and 300 million molecules of hemoglobin, a protein responsible for carrying oxygen throughout the body. A hemoglobin molecule has two subunits, commonly referred to as Alpha and Beta. Both subunits are necessary to bind oxygen and deliver it to cells and tissues in the body. Any condition that affects the quantity or functionality of these components constitute a blood disorder.

Blood disorders have numerous causes and depending on the condition, varying symptoms as well. Some of the common types of disorders that affect the blood include blood cancer (e.g., leukemia), hemoglobinopathies (which affect the structure of hemoglobin molecules in RBCs), clotting disorders and autoimmune disorders. These disorders can be inherited (present at birth) or developed at a later stage in life.

Studies show that every year, 7 million newborns either have a congenital abnormality or a genetic disease. Among the top 5 of these conditions are inherited blood disorders such as Glucose-6-phosphate dehydrogenase deficiency and hemoglobinopathies. Hemoglobinopathies (thalassemia, sickle cell anemia) solely contribute to 330,000 birth defects per year worldwide according to World Health Organization (WHO).

INHERITED BLOOD DISORDERS IN THE MIDDLE EAST

Currently, in the Middle East, data indicates the prevalence rate of hemoglobinopathies to be extremely high. There are several aspects contributing to the increased frequencies and varying molecular characterisation (make-up) of abnormal hemoglobin genes in the region. Consanguineous marriage (inbreeding) practiced as part of the tradition is one of the key factors. Additionally, environmental factors such as history of malaria in the region has enabled carriers of abnormal hemoglobin genes to thrive within the population as it provides immunity against malaria. Other contributing factors are large family size and higher migration rates within the region.
Although population studies are limited and patchy, high disease burden in the Middle Eastern countries warrant introduction of preventive healthcare strategies. Programs such as newborn screening for early detection of inherited blood disorders and premarital counselling for prevention has already been established in Saudi Arabia and the United Arab Emirates. As a result, a dramatic reduction in the number of cases of certain inherited blood disorders (thalassemia) reported has been noted. While preventive healthcare programs like prenatal diagnosis can control further multiplication of the disorders, creating awareness among the public and caregivers is just as important. Such awareness programs may encourage more people to adopt the available resources despite social stigma or cultural restraints. These approaches supported by further research focused on understanding the prevalence pattern and molecular characterisation

of these disorders, can help achieve reduction in the occurrence rate of inherited blood disorders.

The following sections of this booklet cover detailed information about several inherited blood disorders including their symptoms, risk factors, diagnoses, management, and impact on Arab populations. The selection of disorders was done based on disease frequency, accessibility to treatment and availability of screening.