Public Health Practice

Care for people with haemoglobin disorders

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An outline is given of the work of the WHO Collaborating Centre for Haemoglobinopathies, Thalassaemias and Enzymopathies in Saudi Arabia, with special reference to activities related to haemoglobin disorders.

Haemoglobin disorders are a major chronic health problem in several regions of the world; they are particularly common in areas with a history of endemic malaria, where the heterozygous state appears to confer resistance to the growth of the malaria parasite (1).

In Saudi Arabia these disorders occur with high frequency, but the gene frequency and clinical presentation vary from region to region. Among the most common coexisting genes are those of sickle-cell haemoglobin and the α- and β-thalassaemias. During the last two decades, extensive studies have been made of the gene distribution, gene–gene interactions, natural history, complications, management, control and prevention of haemoglobin disorders.

Information for all

In areas where the frequency of the genes is high it is vital that steps be taken to sensitize and inform not only patients and their families but also the general public about what is required in order to tackle the associated problems, for instance those of blood donation, financial support and the employment of people suffering from the disorders. Illustrated talks are given to small groups in hospitals, health care centres and community centres, followed by open discussion of the matters covered. Health care personnel are encouraged to give talks to the families they visit and to underline the significance of screening; furthermore, doctors and other health workers meet informally with patients and their families to discuss such matters. Pamphlets, posters, newspaper articles and television programmes make their contribution, and video films are shown in health centres and outpatient clinics. Information on the disorders is being introduced into school curricula. In order to evaluate the effectiveness of the awareness programme, questionnaires are administered periodically.

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A comprehensive multidisciplinary approach to the care of patients has been adopted. The Sickle-cell Anaemia and Allied Syndrome Study Group (see box), formed in the late 1980s, draws members from a range of specialties, including internal medicine,
paediatrics, clinical pathology, obstetrics and gynaecology, biochemistry and community medicine. The Group is responsible for health care programmes in Riyadh and, through

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local coordinators, elsewhere in the country. At weekly meetings the members discuss the problems of patients and their families, complications, management strategies and other matters. Information is communicated to patients and their families on subjects ranging from home care to the inheritance of the disorders. Patients are followed up regularly; signs, symptoms and complications are recorded, as are the results of haematological, biochemical and radiological investigations. A monograph on guidelines for the management of sickle-cell and thalassaemia disorders has been circulated to doctors in hospitals and clinics with a view to achieving a unified approach nationally.

The national working group, comprising people directly involved with patients suffering from blood genetic disorders, has the functions indicated in the box. The Group is engaged in the following activities.

- Diagnosis, using the facilities at the National Referral and Consulting Unit in Riyadh.
- The application of comprehensive management, therapeutic and care programmes.
- The preparation of regional and national registries.
- Prevention and control through carrier detection and genetic counselling.

**Responsibilities of the Sickle-cell Anaemia and Allied Syndrome Study Group and the National Working Group**

- Multidisciplinary approach to patient care and control of haemoglobin disorders
- Clinical monitoring of individual patients
- Identification and treatment of complications
- Multidisciplinary approach to problems of individual patients
- Monitoring of steady-state and crisis periods (clinical and laboratory)
- Education of patients and families
- Identification of psychosocial problems and their solutions
- Adoption of preventive measures (vaccination, prophylaxis, pharmacological manipulation)
- Screening and genetic counselling

**Creating awareness and educating health care personnel**

- Educational programmes for undergraduate and postgraduate students (medical and paramedical)
- Publication of newsletters
- Seminars, workshops and symposia

**Research**

- Pathophysiology
- New therapies
- Bone marrow transplantation
- Gene therapy
There are regular contacts and consultations, and annual meetings of the national working group are held under the auspices of the Ministry of Health and the College of Medicine at King Khalid Hospital, King Saud University.

Field visits lasting two to three days are conducted by teams of clinicians of the Sickle-cell Anaemia and Allied Syndrome Study Group together with technicians attached to the national referral and consulting unit. The teams hold doctors’ meetings and give lectures with the purpose of updating physicians’ knowledge of haemoglobin disorders; meetings between doctors and patients are arranged; pamphlets are distributed and talks are given to patients and their families in clinics and wards. Wherever possible, genetic counselling is given. Volunteers and suitable patients are included in treatment protocols using hydroxyurea, human erythropoietin, or piracetam.

National meetings are held annually at which experiences and activities are reported, plans for the improvement of health care are discussed, and recommendations are drawn up for the Ministry of Health. Symposia, conferences, training courses and workshops are organized.

Research at King Saud University has led to the application of DNA technology to carrier detection and preclinical and prenatal diagnosis, a major step towards control and prevention. Research is in progress on the management of sickle-cell disease in adults through hydroxyurea therapy and combination therapy involving the use of hydroxyurea and recombinant human erythropoietin, while encouraging results are being obtained in studies on piracetam treatment in children.

Policy-makers and administrators need to know the magnitude of the problems, the measures necessary for prevention and control, and how such measures can be cost-effectively incorporated into primary health care; since they also have to be made aware of the merits of proposals, evaluations have to be carried out.

**Regional and university activities**

In 1991, the World Health Organization designated the Department of Medical Biochemistry in King Saud University as the WHO Collaborating Centre for Haemoglobinopathies, Thalassaemias and Enzymopathies. The Centre’s main responsibilities are:

- improvement of the delivery of thalassaemia control services to the community;
- investigation of community knowledge and attitudes to the control of hereditary diseases;
- development of training aids for the hereditary diseases control programme;
- assistance with training in population screening and fetal diagnosis of haemoglobin disorders;
- development and improvement of diagnostic methods for haemoglobin disorders and enzymopathies;
- investigation of the structural, functional and genetic aspects of haemoglobin disorders and enzymopathies.

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It should be noted that health care at the national level is linked to that at the regional (i.e., countries in the Eastern Mediterranean) level, which in turn is linked to that at the international level (see figure). The Centre
National, regional and international links

The control of haemoglobin disorders can be achieved only through multifaceted programmes directed towards families and the communities where affected individuals live. Adequate plans and resources are essential, as is the coordination of the activities of administrative, clinical and scientific bodies.

A heightened awareness among families and communities on the one hand and clinicians and paramedical personnel on the other can favourably alter attitudes towards control programmes. Good global control requires cooperation, close monitoring, learning from each other’s experience, and the implementation of programmes in as unified a manner as possible.

Reference