Septicaemia in Sickle Cell Disease Patients at Qatif Central Hospital, Saudi Arabia

Sir,

I have read the article by A. M. El-Bashier et al. (Saudi Med J 1992; 13(3): 220–223) on septicaemia in sickle cell disease patients (SCD) at Qatif Central Hospital. The topic of SCD in Saudi Arabia has attracted worldwide interest resulting in several studies and publications. It is unfortunate that the authors did not refer to more recent reports which are more relevant to the subject of their study. In this regard, I would like to make the following comments:

1. It is known that in Saudi Arabia there are at least two types of SCD which are distinguishable clinically, haematologically, and genetically. The 'mild' type occurs predominantly in patients whose ancestral origin is from the Eastern Province no matter where they live and is similar to descriptions from certain regions of India. The more severe type occurs predominantly in patients whose ancestral origin is in the southwest of the Arabian peninsula, no matter where they live, and is similar to the disease described in Western literature in patients of African ancestry.

2. The risk for infections is different in these two groups of patients. In a prospective controlled study of infections in early childhood in patients with SCD, who were born and lived in the Eastern Province, we found that patients of eastern origin have no increased risk for infections in general. This finding contrasts with the high risk of overwhelming infections in young children of southwestern origins.

3. It would have been more informative if the authors had reported the ancestral origins of their patients in particular the children who died of overwhelming infections. In addition, differentiating between early childhood (up to 5 years) and later childhood, would lead to better definition of age at risk for infections and to results that are comparable with the literature. These age and origin risk factors for infections (as well as for other clinical features of SCD) are important to identify because of prophylactic and therapeutic implications.

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References


Sir,

We would like to thank Professor El-Mouzan for bringing to our attention some of the interesting findings of other groups and for his suggestions especially with regards to age breakdown for analysis.

We wish to state, however, that our limited objectives are as stated in the second paragraph of our article, to identify causative organisms in septicaemia, their antibiotics susceptibility and their mortality rate. However, in previous studies we have clearly demonstrated that the disease is not as benign as was thought.

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References


Psychiatric Implications of the Wolfram Syndrome Gene

Sir,

I read with great interest the review of the Wolfram syndrome and the detailed case history by Drs Rifat Naghmi and his associates. From a medical point of view this review paper described all the most clinically significant details of Wolfram syndrome. In addition, thiamine-dependent sideroblastic anaemia has recently been reported in two paediatric patients from the Arab World. However, from a psychological perspective these papers did not highlight at all the psychiatric disorders in the families of Wolfram syndrome recently reported in two well designed researches. This might be
because the patients or the relatives did not manifest any psychiatric symptoms or because the authors did not look for them. However, the following comment which is mainly extracted from two articles might caution the clinicians to search for psychiatric symptomatology in the probands or in the family members of Wolfram syndrome patients and, accordingly, refer such patients to psychiatric liaison services for comprehensive medical as well as mental health care.

The DIDMOAD (Wolfram) syndrome is a neurodegenerative classical Mendelian autosomal recessive syndrome (ARS) which is mainly characterized by diabetes mellitus, progressive optic atrophy diabetes insipidus, deafness, an atonic bladder, diverse neurological abnormalities and is rarely associated with thiamine dependent anaemia. Generally speaking there are about 0.5-1% heterozygous carriers of any single autosomal recessive syndrome in the general population.

However, this figure for homozygous carriers varies from 1 in 40 000 to 1 in 160 000. The well defined homozygous carriers of a gene for autosomal recessive syndrome have a very high rate of developing a common chronic disease. On the other hand the heterozygous carriers of a specific gene also have a high risk of a common disease. The important implication of this is that the specific gene could also predispose members of the general population to the common chronic disease.

Psychopathology has been described in 60% of patients who were homozygous for Wolfram syndrome. The reported psychiatric manifestations were depression, violent or assaultive behaviour, and organic brain syndrome. A high prevalence of these psychiatric symptoms has also been found in mentally sick relatives of patients with the Wolfram syndrome. This might reflect that these specific psychiatric symptoms may also be associated with heterozygosity for the Wolfram syndrome gene. Therefore, it has been hypothesized that heterozygous carriers of the gene for the Wolfram syndrome may be predisposed to significant psychiatric illness. This hypothesis has been tested by a controlled study and encouraging results were found. In this research 36 families with Wolfram syndrome probands were identified and the sample comprised of psychiatrically ill blood relatives (n = 543) and their spouses (n = 398). The data from their families were compared with those obtained in blood relatives (n = 1595) and their spouses (n = 1100) from 110 families ascertained by means of ataxia-telangiectasia probands.

Ataxia-telangiectasia, like the Wolfram syndrome is an autosomal recessive syndrome with similar high morbidity in adolescence and early adult life. The homozygous individual with ataxia-telangiectasia develops progressive cerebellar ataxia and other neurological symptoms and dies young from breast cancer or progressive pulmonary disease.

As a result of these comparisons the following significant findings were observed.

A significantly larger proportion of blood relatives than spouses in the families with Wolfram syndrome had undergone psychiatric hospitalizations, or had committed suicide or had self-reported mental illness or chronic nervous trouble, the four important clinical parameters unaffected by diagnostic inconsistencies.

The proportions of blood relatives with evidence of psychiatric illnesses were very similar to those of the spouses in the ataxia-telangiectasia families.

The similar four clinical indices were significantly more common in blood relatives in families with Wolfram syndrome than in blood relatives in families with ataxia telangiectasia.

No statistically significant differences were found between the spouses in the two groups of families (p > 0.05).

On the basis of these findings the following conclusions were drawn:

1. Individuals heterozygous for the Wolfram syndrome are predisposed to psychiatric disorders.

2. Individuals heterozygous for the Wolfram gene which represent 1% of the general population might account for almost 8% of all psychiatric admissions or suicides in the USA.

3. The Wolfram syndrome is an autosomal recessive syndrome [MIM222300]. If the gene responsible for this syndrome can be mapped and cloned by using molecular genetic techniques, then the range of specific psychopathology associated with this gene can be distinctively defined in families with the Wolfram syndrome.

4. Understanding the metabolic action of the Wolfram syndrome gene which is until now elusive will lead to primary effective prevention or treatment of the associated psychiatric illness.

In conclusion, besides being associated with multiple chronic medical diseases, the homozygous and heterozygous carriers of the Wolfram syndrome gene also manifest certain psychiatric features. Therefore all clinicians should screen the entire family of individuals with the Wolfram syndrome for psychopathological entities.

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References


Sirs,

We appreciate the descriptions of psychiatric disorders in families with Wolfram syndrome as quoted by Dr Qureshi. Our patient and his family members did not have any evidence of psychiatric illness. Conversely, we found the reported case to be rather docile and with a placid temperament. Coincidentally at the moment one of the sisters of the reported patient is undergoing hospitalization and has been diagnosed to have DIDMOAD syndrome. She too lacks any psychiatric illness.

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