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FOCUS ON PRACTICE

What are the implications of sickle cell anaemia?

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Sickle Cell Trait is common in certain populations. The vast majority of individuals who possess combinations of these genes which cause variations in haemoglobin are asymptomatic.

However, despite advances in treatment, health problems associated with homozygous inheritance can result in work-related performance issues. Such cases require individual risk assessments in relation to their jobs when considering fitness for work, rehabilitation and prevention of symptoms.

Key words: Employment policy; occupational health; sickle cell anaemia.

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INTRODUCTION

The reduced solubility of Haemoglobin S, which in the deoxygenated state results in the occurrence of aggregates of sickled haemoglobin, accounts for the clinical features of sickle cell anaemia. There are several disorders involving sickle haemoglobin. These are the heterozygous state for haemoglobin S, or the sickle cell trait (AS); the homozygous state, or sickle cell disease (SS); and the compound heterozygous state for haemoglobin S together with haemoglobin C, D, E or other structural variants.

There is considerable variation with which different haemoglobins are able to interact with haemoglobin S and this accounts for some of the clinical variability of the different sickling conditions.

The disorders occur very frequently in African Black populations and sporadically throughout the Mediterranean region and the Middle East. The disorder has not been seen in South East Asia but there are extensive pockets of the disorder in India.

In practice AS is asymptomatic and causes no haematological changes. The evidence for problems in AS individuals in occupational groups following extreme anoxia is sparse.[1]

Sickle cell anaemia is produced by the homozygous inheritance of the sickle gene which has a variable clinical expression. This ranges from a severe haemolytic anaemia associated with painful crises to a mild anaemia found on routine screening.

Severe cases present in infancy and although growth and development are usually normal, skeletal deformities (*e.g.* frontal bossing) and evidence of chronic leg ulceration should alert one to the possibility of sickle cell disease. Severe cases result in individuals being unable to gain skills or qualifications owing to the frequency of medical complications.

The complications of sickle cell anaemia result in several occupational health issues. Sickle cell crisis can be precipitated by dehydration, exposure to cold, infection and environments with low oxygen tension, *e.g.* work at high altitudes. As a result, occupations such as commercial divers and aircrew are considered unsuitable for individuals with the SS trait. Individuals with AS are also frequently restricted from military aircrew roles, although the evidence showing problems associated with AS is lacking in practise. Serious forms of thrombotic crisis — 'lung' and 'brain' syndromes, are characterised by acute dyspnoea, pleuritic pain and episodes resembling transient ischaemic attacks.[2] These could result in significant incapacitation during work activities.

While some individuals with sickle cell anaemia manage well with relatively low haemoglobin, it would be reasonable to avoid occupations, where potential exposure to substances such as lead and amino and nitro compounds could exacerbate the chronic haemolysis;[3] however, the risks may have been exaggerated.[4]

While many individuals with SS can go though life with few complications, there is a subset who undergo repeated crisis, neurological complications and incapacitating bone disease. With the success of several treatment initiatives such as hypertransfusion, bone marrow transplantation, hydroxyurea treatment raising haemoglobin F which protects against sickling, prophylactic antibiotics and pneumoccal vaccine, the course and prognosis of sickle cell disease has improved.

From an employment perspective absence from work either for treatment or complications is an issue. The nature of the work, the risks involved, the potential to

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make reasonable adjustments in order to avoid discriminatory issues[5] and the likelihood of complications need to be assessed before any decision is made regarding suitability for employment. Past history from the GP or Specialist such as frequency of crisis (severe > 3/year), genotype (SC better prognosis than SS), should form part of the risk assessment.

The presentation of complications of SS can give rise to diagnostic challenges to an occupational health department. The painful crisis gives rise initially to vague pain but can progress to severe pains with a bizarre distribution. A thrombotic crisis can manifest in one or several systems and sequestration can lead to serious and multiple infections.

Frequent episodes of in-patient treatment or therapy can result in psychological problems resulting in prejudice and a lack of tolerance in work colleagues and from management.

Chronic complaints are usually a result of repeated episodes of vascular occlusions. Vision may become impaired with a proliferative retinopathy and chronic pulmonary fibrosis is increasingly being recognized as a cause of respiratory complications.

In terms of prevention, avoidance of cold temperatures, excessive exertion and dehydration whilst at work is simple but practical advice.

While many SS individuals are employed in a wide variety of jobs, unemployment is high (> 70%) in such groups, in part reflecting the course and complications of the condition.

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