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SHORT COMMUNICATION

Arthropathy in sickle cell disease

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Acute joint pain, with swelling and limitation of motion, has been reported in individuals with either homozygous or heterozygous sickle cell disease (Orozco-Alcala & Baum, 1973; Saheb, 1973).

Sickling of red cells in small periarticular blood vessels, with subsequent thrombosis and ischaemia, are thought to be the cause (Schumacher, Andrews & McLaughlin, 1973). The differential diagnosis of atypical joint manifestations, even when limited to hereditary disease, is a lengthy one (Kats, 1965). Sickle cell arthropathy may so far not have been considered as a cause of sudden joint pain occurring during flight. Commercial aircraft flying above 24,000 ft altitude, pressurize their cabin altitudes between 5000 and 7000 ft. Thrombo-embolic events in individuals with the sickle cell trait have been reported at altitudes lower than that (Kats, 1973a). Although the risk of vaso-occlusive events, occurring in flight, may be very small, it is worth while to keep the diagnosis in mind. Presently, there exist no objective measurements, that allow us to predict, which carrier of the trait is at risk (Kats, 1973b).

The diagnosis of acute sickle cell arthropathy must be differentiated from another type of acute joint pain that may develop in flight. With reduction of barometric pressure, nitrogen, dissolved in the body fluid and fat compartments, evolves in the form of gas bubbles (Billings, 1973). These bubbles may produce pain, especially around the joints. These joint pains are generally referred to as 'the bends'. They are a potential problem in divers who ascend to the surface of the water after minutes or hours at depth, and also in pilots who climb in their aircraft rapidly to altitude. In pilots the bends usually occur above 18,000 ft (barometric pressure 380 mmHg), but scuba diving in the previous 24 h may lower this threshold to 8000 ft (565 mmHg). Previous exercise, overweight, old age and decrease in environmental temperature may lower this threshold even further. During World War II and the Korean crisis, screening tests for sickle cell disease were not performed routinely for selection of military flight personnel. Africans at that time served in the R.A.F. and investigations were carried out in West Africa (Findlay *et al.* 1947) to determine in human volunteers, the factors which precipitate acute sickling. It is quite possible, that among pilots, who suffered then from the bends, some were actually sickle trait carriers with arthropathy.

West Africa, especially Ghana, has a high prevalence of haemoglobin SC disease, and to a lesser extent of haemoglobin AS disease (Kats, 1972). Central Africa, with, as in Zambia (Kats, 1973c), plateaus up to 7000 ft, has a lower prevalence of these haemoglobinopathies. It can be speculated, that altitude has constituted a selective force, detrimental to the trait carrier, and that therefore the prevalence in Zambia is lower. Research in this areas is timely, and at the same an increased awareness among physicians and aircraft cabin attendants is needed, to identify those signs and symptoms, that may be attributed to underlying haemoglobinopathies.

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References

Billings, C.E. (1973) *Bioastronautics Data Book*, 2nd edn, p. 25. N.A.S.A.

Findlay, G.M., Boulter E.A. & MacGibbon C.B. (1947). A note on sickling and flying. *J. R. Army med. Cps*, **89**, 138–141.

Kats, B.A. (1965) Atypical joint manifestations in a patient with familial mediterranean fever. *Ned. T. Geneesk*, **109**, 373–376.

Kats, B.A. (1972) Ghana, aviation and medicine. *Flying Physician*, **15**, 34–35.

Kats, B.A. (1973a), Risk in sickle-cell trait. *Ann. Intern. Med.* **79**, 289–290.

KATS, B.A. (1973b) Search for a 'sickling index'. *Lancet*, ii, 742–743.
KATS, B.A. (1973c) Zambia, aviation and medicine. *Flying Physician*, **16**, 26–27.
OROZCO-ALCALA, J. & BAUM, J. (1973) Arthritis during the sickle cell crisis. *N. Engl. J. Med.* **282**, 420.
SAHEB, F. (1973) Acute joint pain in a patient with haemoglobin SC disease. Case report. *N. Engl. J. Med.* **288**, 971.
SCHUMACHER, H.R., ANDREWS, R., MCLAUGHLIN, G. (1973) Arthropathy in sickle-cell disease. *Ann. Intern. Med.* **78**, 203–211.

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