

should be preventable in many patients at particular risk of this unpleasant problem.

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- ¹ Neviasa JS. Adhesive capsulitis of the shoulder. A study of the pathological findings in peri-arthritis of the shoulder. *J Bone Joint Surg* 1945;**27**:211-22.
- ² Bruckner FE, Nye CJS. A prospective study of adhesive capsulitis of the shoulder ("frozen shoulder") in a high risk population. *Q J Med* 1981; **198**:191-204.
- ³ Wright V, Haq AMMM. Peri-arthritis of the shoulder. II. Radiological features. *Ann Rheum Dis* 1976;**35**:220-6.
- ⁴ Reeves B. Arthrographic changes in frozen and post-traumatic stiff shoulders. *Proceedings of the Royal Society of Medicine* 1966;**59**:827-30.
- ⁵ Hazleman BL. The painful stiff shoulder. *Rheumatology and Physical Medicine* 1972;**11**:413-21.
- ⁶ Wright V, Haq AMMM. Peri-arthritis of the shoulder. I. Aetiological considerations with particular reference to personality factors. *Ann Rheum Dis* 1976;**35**:213-9.
- ⁷ Van der Korst JK, Colenbrander H, Cats A. Phenobarbital and the shoulder-hand syndrome. *Ann Rheum Dis* 1966;**25**:553-5.
- ⁸ Johnson JTH. Frozen shoulder syndrome in patients with pulmonary tuberculosis. *J Bone Joint Surg* 1959;**41A**:877-82.
- ⁹ Bridgman JF. Peri-arthritis of the shoulder and diabetes mellitus. *Ann Rheum Dis* 1972;**31**:69-71.
- ¹⁰ Rathbun JB, MacNab I. The microvascular pattern of the rotator cuff. *J Bone Joint Surg* 1970;**52B**:540-53.
- ¹¹ McNab I. Rotator cuff tendinitis. *Ann R Coll Surg Engl* 1973;**53**:271-87.
- ¹² Bulgen D, Hazleman B, Ward M, McCallum M. Immunological studies in frozen shoulder. *Ann Rheum Dis* 1978;**37**:135-8.
- ¹³ Bulgen DY, Hazleman BL, Voak D. HLA-B27 and frozen shoulder. *Lancet* 1976; *ii*:1042-4.
- ¹⁴ Coventry MB. Problem of painful shoulder. *JAMA* 1953;**151**:177-85.
- ¹⁵ Fleming A, Dodman S, Beer TC, Crown S. Personality in frozen shoulder. *Ann Rheum Dis* 1976;**35**:456-7.
- ¹⁶ Lloyd-Roberts GC, French PR. Peri-arthritis of the shoulder. A study of the disease and its treatment. *Br Med J* 1959; *ii*:1569-71.
- ¹⁷ Thomas D, Williams RA, Smith DS. The frozen shoulder: a review of manipulative treatment. *Rheumatol Rehabil* 1980;**19**:173-9.
- ¹⁸ Quin CE. Humeroscapular peri-arthritis. Observations on the effects of x-ray therapy and ultrasonic therapy in cases of "frozen shoulder." *Annals of Physical Medicine* 1969;**10**:64-9.
- ¹⁹ Williams NE, Seifert MH, Cuddigan JHP, Wise RA. Treatment of capsulitis of the shoulder. *Rheumatol Rehabil* 1975;**14**:236.

Indian childhood cirrhosis

A unique liver disease, which has resisted all attempts at understanding, affects young children in the Indian sub-continent and Malaysia. It is said to be extremely rare among expatriate children; information about the number of cases encountered in Britain (or in other parts of the world where Indians have settled) would be valuable. The disease is not confined to any one religious group or social class (a preponderance of cases from the upper classes is due to their better access to medical care), and in a third of cases it affects more than one member of the family.

In most of the children¹ non-specific symptoms, abdominal distension, and enlargement of the liver are succeeded in a few years by death from decompensated cirrhosis, causing either liver failure, ascites, or bleeding from portal hypertension. In a quarter the illness is more acute and resembles a continuing

hepatitis, with death from liver failure in a few months. The liver damage is striking and unusual. The characteristic features are appreciable necrosis of liver cells with little sign of regeneration, extensive deposition of hyaline (especially in liver cells), patchy and "aggressive" fibrosis throughout the parenchyma, very little fat, and a scanty inflammatory cell response. The resulting damage has been called "micro-micronodular" cirrhosis.

The familial incidence suggests a genetic susceptibility to one or more environmental hazards, but no definite cause has been recognised. Malnutrition is clearly not a candidate, though some dietary substance or deficiency peculiar to a particular culture might still be responsible. Cell-mediated immunity is depressed in some patients and on the basis of raised concentrations of alpha-fetoprotein in nearly half their cases, Nayak *et al*² made the ingenious suggestion that persistence of fetal hepatocytes made the children vulnerable to an environmental agent. One of the hepatitis viruses is an obvious choice. The frequency of hepatitis B surface antigen, however, though high, is not remarkable for these populations, and no sign of hepatitis B has been found in biopsy or necropsy specimens of liver tissue.³ Ingestion of a hepatotoxin such as aflatoxin seems to be disproved by the histological appearances, but perhaps ayurvedic remedies should not be entirely ruled out, since drugs like griseofulvin and colchicine cause hyaline deposits in the liver of animals.

Recent interest has centred on the finding of large amounts of copper and copper-binding protein in the liver.⁴⁻⁶ This feature, with the hyaline deposits, is reminiscent of Wilson's disease. Whether these appearances are primarily metabolic or the consequence of the disease—as in prolonged cholestasis, for example—is not yet clear, though their distribution in liver cells favours a causative role. But careful study⁷ of siblings of affected children has shown only minor "non-specific" changes in liver biopsy material and no excess of copper, and prolonged follow-up of 200 siblings has not produced a single case of Indian childhood cirrhosis. Obviously the next step is a controlled trial of penicillamine. Nevertheless, while we naturally hope that children will benefit, use of the drug may not throw any new light on this curious disease.

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- ¹ Nayak NC, Ramalingaswami V. Indian childhood cirrhosis. *Clin Gastroenterol* 1975;**4**:333-49.
- ² Nayak NC, Chawla V, Malaviya AN, Chandra RK. α -Fetoprotein in Indian childhood cirrhosis. *Lancet* 1972; *ii*:68-9.
- ³ Nayak NC, Ramalingaswami V, Roy S, Sachdeva R. Hepatitis-B virus and Indian childhood cirrhosis. *Lancet* 1975; *ii*:109-11.
- ⁴ Portmann B, Mowat AP, Tanner MS, Williams R. Orcein positive liver deposits in Indian childhood cirrhosis. *Lancet* 1978; *ii*:1338-40.
- ⁵ Tanner MS, Portmann B, Mowat AP, *et al*. Increased hepatic copper concentrations in Indian childhood cirrhosis. *Lancet* 1979; *ii*:1203-5.
- ⁶ Popper H, Goldfischer S, Sternlieb I, Nayak NC, Madhavan TV. Cytoplasmic copper and its toxic effects. Studies in Indian childhood cirrhosis. *Lancet* 1979; *ii*:1205-8.
- ⁷ Nayak NC, Marwaha N, Kalva V, Roy S, Ghai OP. The liver in siblings of patients with Indian childhood cirrhosis: a light and electron microscopic study. *Gut* 1981;**22**:295-300.