

will advise the Secretary of State for Health and Social Services and the Secretary of State for Wales on co-ordinating and developing health and personal social services as they relate to children and families with children. At the first meeting the committee asserted that our society had the potential to build child care and health services as good as any in the world; but to attain that objective we will need to give a higher priority to improvements in these services. The new committee will welcome advice from all statutory and voluntary agencies and from individuals, and intends to find out both why services are working well in some areas and why they are less successful in others. Then it plans to use its power of direct access to the Secretaries of State to press for practical measures and to influence public opinion.

With these fine sentiments the Children's Joint Committee has been given three years in the first instance to justify its existence. At least one facet of the Court Report has been implemented. We wish the committee success.

<sup>1</sup> Committee on Child Health Services, *Fit for the Future*, volumes 1 and 2. London, HMSO, 1976. (Court Report.)

## Behçet's disease

For a disease of such rarity Behçet's disease enters into differential diagnosis quite often. In part this is because it may attack so many different organs: any combination of bizarre symptoms and signs together with mouth or genital ulcers should bring Behçet's disease to mind. Even so, it is rare for a doctor practising in Britain to diagnose more than one case in a lifetime. Rheumatologists perhaps see more than their fair share—largely because any puzzling disorder with ocular lesions and associated with joint disease gravitates towards them.

The classical triad of relapsing iritis with ulcers of the mouth and genitalia named after Behçet<sup>1</sup> was first described by Blüthe in 1908.<sup>2</sup> Since then, additional features have been described and shown to be clearly included in the variable pattern of the disease. In a recent series of 41 patients<sup>3</sup> the main site was the eye in 10 patients, the joints in eight, phlebitis often affecting the venae cavae in eight, and mucocutaneous or mucous lesions in nine; while neurological defects were the predominant manifestation in six patients, and erythema nodosum in one. This catalogue of systems was reflected in the clinics at which these patients were first seen: ophthalmological in 11; medical in 10; dermatological in nine; neurological and vascular three each; oral two; and gynaecological, urological, and ear, nose, and throat one each. The diagnostic clue was provided by oral ulceration, which was the earliest manifestation in 30 cases, was the presenting feature in 11, and occurred at some time in all but one. Genital ulceration occurred in 36 of the 41 patients. Another classical diagnostic clue, blister formation or an inflammatory reaction at a venepuncture or scratch site, occurred in only 15.

Behçet's syndrome relies on suspicion and clinical acumen

alone for diagnosis, but a novel approach has come from Leeds.<sup>4</sup> Behçet's disease, together with Whipple's disease, has been firmly placed in the group of spondarthroses<sup>5</sup> seronegative for rheumatoid factor on account of the linking or overlap of its principal features (peripheral joint lesions, sacroileitis, buccal ulceration, erythema nodosum, and thrombophlebitis) with other more established members of this group, including ulcerative colitis, Crohn's disease, ankylosing spondylitis, psoriatic arthritis, and Reiter's disease. Familial associations of the more common diseases in this group are now well recognised, suggesting a genetic component; but Behçet's and Whipple's diseases are so rare that no such association has yet become apparent.

Chamberlain<sup>4</sup> did a survey in Yorkshire looking for cases of Behçet's disease with the help of general practitioners and hospitals and learnt of 41 cases in a population of five million. Of the 33 patients seen, 32 fulfilled her diagnostic criteria. The survey might have been expected to yield some cases with a more benign pattern: in fact, it confirmed the potential serious nature of the disease. Symptoms developed almost always between the ages of 15 to 40. One patient died with lesions affecting the central nervous system. Ocular damage was milder in the Yorkshire patients than in those studied in the Levant, where posterior segment lesions caused total loss of vision, usually in both eyes, in eight of 41 patients.<sup>3</sup>

Clues to aetiology are few and far between. Reports of clustering, particularly from Turkey and Japan, suggest the relevance of infective or environmental factors; while studies from Israel, where the disease occurs (along with multiple sclerosis) more commonly in immigrants than in the same groups in their countries of origin, are against a heritable disease of low penetrance.<sup>3</sup> At a recent symposium on Behçet's disease in Istanbul environmental pollutants such as organochlorine compounds, pesticides, and copper were put forward as aetiological factors or triggers; thus guinea pigs receiving these chemicals for a year had developed oral aphthae, pyoderma, and genital ulcers in contrast to controls.<sup>6</sup> Some support for a genetic component, comparable with that clearly shown in ankylosing spondylitis, comes from the Leeds survey, in which a modestly increased frequency of HLA B5 and B27 was found, confirming a report of increased frequency of B5 in Japan.<sup>7</sup> HLA B5 also appears more frequently in those patients with uveitis and Behçet's disease.<sup>8</sup>

There has been little progress in the treatment of Behçet's disease. Topical corticosteroids may be of value in the eye and mouth, while systemic corticosteroids, immunomodulatory drugs such as levamisole and penicillamine, and immunosuppressive drugs have also been claimed to be of benefit. Transfer factor was once thought to be of value, but the results of a recent controlled study have failed to confirm this. On the whole, the rarity of the disease means that most treatment will be empirical.

<sup>1</sup> Behçet, H, *Dermatologische Wochenschrift*, 1937, **105**, 1152.

<sup>2</sup> Blüthe, L, *Inaugural Thesis*, Heidelberg 1908.

<sup>3</sup> Chajek, T, and Fainaru, M, *Medicine (Baltimore)*, 1975, **54**, 179.

<sup>4</sup> Chamberlain, M A, *Annals of the Rheumatic Diseases*, 1977, **36**, 491.

<sup>5</sup> Moll, J M H, et al, *Medicine (Baltimore)*, 1974, **53**, 343.

<sup>6</sup> Ishikawa, S, *International Symposium on Behçet's Disease*, Istanbul, 1977.

<sup>7</sup> Ohno, S, et al, *Lancet*, 1973, **2**, 1383.

<sup>8</sup> Hamza, M, *International Symposium on Behçet's Disease*, Istanbul, 1977.