eventually died from multiorgan failure. Oral anticoagulation probably prevented the development of major arterial and venous thromboembolic disease, without causing the rupture of any of the pre-existing aneurysms. However, anticoagulation did not prevent the progression to multiorgan failure, which can be attributed to alterations in the microvascular circulation.

In conclusion, we think that multiple splachnic aneurysms probably represent part of the spectrum of vascular abnormalities of primary APS. If such aneurysms are identified, lifetime anticoagulation should still be considered as the preferred treatment in order to prevent deep venous thrombosis and/or pulmonary embolism, despite the risk of bleeding complications.

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A family with diffuse idiopathic skeletal hyperostosis

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We report a family with diffuse idiopathic skeletal hyperostosis (DISH). The most striking occurrence was severe cervical disease without extensive dorsal involvement. From the tissue typing results of our two sibling patients, it appears less likely that, if there is a hereditary component, it is linked to HLA status. It remains to be seen whether this is a new disease entity or an unusual familial variant of DISH. We are unaware of a similar published report.

A 23 year old man was referred with a painful stiff neck of 3 years' duration. On examination, all movements of his cervical spine were restricted. Other spinal movements were normal. Inflammatory markers were normal and HLA-B27 was negative. An *x* ray examination of the sacroiliac joints, thoracic and lumbar spine were unremarkable. However, the cervical spine radiograph showed gross anterior osteophytosis (fig 1A).

This man's 24 year old sister had been seen 7 years previously. She described a 6 year history of worsening neck pain and stiffness. On examination, movements of the cervical spine were severely limited in all directions, with mild limitation of the thoracic spine. An *x* ray examination and inflammatory markers were normal and HLA-B27 was negative. Five years later, the clinical findings had scarcely changed. However, although the sacroiliac joints were still normal, there was now marked osteophytosis around the hip joints with gross osteophytosis and ankylosis of the cervical spine (fig 1B). Two years later, this advanced cervical pathology precipitated cervical myelopathy.

The father of these patients was first seen at 52 years of age despite having a ''30 year history of ankylosing spondylitis''. He had a strong family history of the disease, with brother, sister, and mother affected. On examination, all spinal movements were markedly reduced. Movements of both hips were severely restricted and bilateral elbow fixed flexion deformities were present. Inflammatory markers were normal and HLA-B27 was negative. An *x* ray examination showed normal sacroiliac joints, advanced osteophytosis and degeneration of the hip joints, and ankylosis of the cervical spine (fig 1C). His hip disease required prompt replacement surgery.

All these patients had radiological changes suggestive of DISH. Two of this patient's siblings also had the disease, as do his other two offspring (they are receiving care at different hospitals). The first two patients described here were tissue typed: these siblings only shared alleles at DRB3 and the C locus, which occur frequently in the general population.

DISH is an ossifying, non-inflammatory, non-erosive enthesopathy favouring the dorsal spine but sparing the sacroiliac joints. By contrast, ankylosing spondylitis is an inflammatory condition with enthesopathies facing joints, always affecting the sacroiliac joints. DISH affects 3–6% of the population over 40 years of age and 11% aged over 70 years.[1] It is twice as common in men and occurs more frequently in certain racial groups: it is common in Japanese and Pima Indians but rare in black and Asian races.[1] Other causes of hyperostosis or bony excrescences include spondylitis deformans, ankylosing spondylitis, trauma, fluorosis, treatment with retinoids, ochronosis, acromegaly, hypoparathyroidism, and x-linked hypophosphataemic osteomalacia, but there was nothing in the history, physical examination, or the investigations to suggest that our three patients had any of those conditions.[2]

In the cervical spine, ossification of the posterior longitudinal ligament (OPLL) is commonly seen. This phenomenon is often called ''Japanese disease'' owing to its predominance in the Japanese population.[3] OPLL displays a strong genetic component with high concordance in twins and families.[4 5] Various modes of inheritance have been suggested, including HLA linkage.[6] In DISH, however, although there are racial differences, no strong familial tendency has been demonstrated. Neither is there a proven

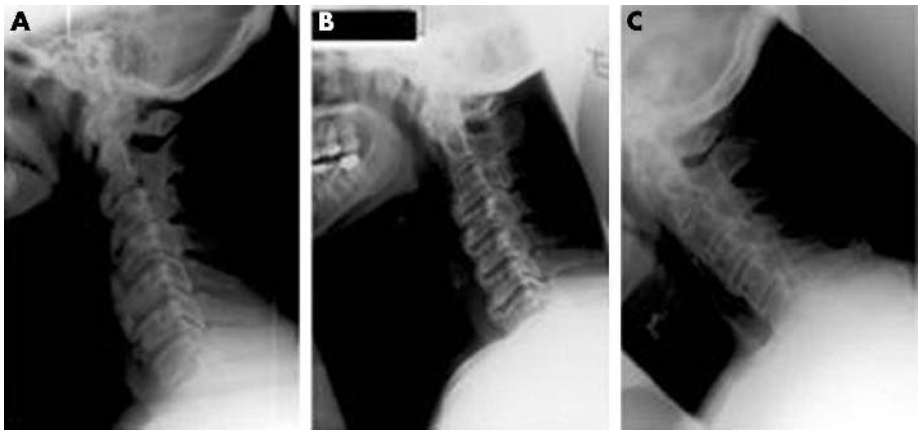

Figure 1 Lateral *x* ray findings of the cervical spine of the son (A), the daughter (B), and the father (C), showing severe anterior osteophytosis.

HLA link, as found between HLA-B27 and the spondyloarthropathies.

Although our patients were diagnosed as DISH, there are some atypical features. The most striking of these is severe cervical disease without extensive dorsal involvement. Forestier and Rotes-Querol in their classification criteria, considered involvement of at least three intervertebral bodies in the dorsal spine to be essential.[7] However, Utsinger's criteria do not include this as a necessary feature.[8] One similar case has been described: a 71 year old patient presented with similar cervical findings and sparing of the dorsal spine and sacroiliac joints. Difficulty was found classifying the condition as either DISH or ankylosing spondylitis.[9] Another unusual feature in our cases is the strong familial pattern. From the tissue typing results of our two sibling patients, it appears less likely that, if there is a hereditary component, it is linked to HLA status.

It thus remains to be seen whether this is a new disease entity or an unusual familial variant of DISH.

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Antiphospholipid antibodies in patients with scleroderma: prevalence and clinical significance

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Antiphospholipid antibodies (aPL) are detected in a variety of autoimmune disorders, most commonly systemic lupus erythematosus, but also in some infectious diseases, lymphoproliferative disorders, and even in apparently healthy people.

Although a wide prevalence of aPL in systemic sclerosis has been reported (between 0 and 41%), most studies have focused on anticardiolipin antibodies (aCL) and very little is known about other aPL in this disease. We determined the prevalence and clinical significance of aCL, antibodies to β_2-glycoprotein I (anti-β_2GPI), and antibodies to phosphatidylserine-prothrombin complex (aPS-PT) in 25 patients with scleroderma (18 with limited and 7 with diffuse scleroderma, as defined by LeRoy *et al*[1]) (table 1). Twenty four patients were female (median age 50 years (range 28–70), median disease duration 3 years (range 1–20)). One patient had a history of venous thrombosis. Of the 17 patients who had ever been pregnant, five had an adverse obstetric history. Two patients had miscarriages (before the 10th week of gestation), two patients had a fetal death (at the 10th week of