Sturge–Weber syndrome

Sir,

The cutaneous manifestations of Sturge–Weber syndrome have been described by Romanowski and Cavallin (Vol 59(3), 1998, p. 226), in turn, as a facial cutaneous vascular naevus, a naevus flameus, a port wine naevus and also as a capillary angioma of the dermis. Such terminology is confusing to paediatricians, plastic surgeons and dermatologists dealing with vascular anomalies who are interested in their clinical course, prognosis and efficacy of different modalities of treatment.

The understanding of vascular anomalies has been greatly facilitated by Mulliken and Glowacki (1982) who proposed the biological classification system dividing them into those that are biologically active (haemangiomas) and those that are biologically inert (vascular malformations).

This system has gained wide acceptance by clinicians dealing with such problems as it is easy to understand and explain to patients and also provides a rational basis from which to formulate an appropriate therapeutic plan.

The cutaneous manifestation of Sturge–Weber syndrome, described as a port wine stain, is an intradermal capillary malformation. This vascular malformation is present at birth, persists throughout life, growing proportionately with the patient, and does not undergo regression. It is not a haemangioma as it is not a biologically active endothelial tumour that exhibits a distinct proliferative phase followed by slow involution phase. It cannot also be described as a naevus.

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Mulliken JB, Glowacki J (1982) Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. *Plast Reconstr Surg* **69:** 412–20

Pressure sores and severe sepsis

Sir,

The case study by Kyriakides et al (Vol 59(11), 1998, p. 896) is an excellent illustration of the acute susceptability to peripheral tissue necrosis as a result of pressure caused by severe sepsis discussed in my article in the same issue (Vol 59(11), 1998, p. 841). It also demonstrates the value of providing an effective pressure relieving support in the immediate postoperative period for this very sick young woman. The only part of the body which developed a pressure sore was the occiput which was presumably resting on a pillow. The authors used a low air-loss bed, but alternating pressure air mattress overlays, which are cheaper and easier to install, have also been shown to prevent pressure injuries in intensive care patients (Gebhardt et al, 1996).

As this patient's pressure sore was not noted until the thirtieth postoperative day, the injury was unlikely to have occurred on the operating table, but this may happen, as the authors point out. They discuss the importance of repositioning the head every 30 minutes to prevent occipital sores and alopecia in patients undergoing prolonged surgery. However, they do not mention the rest of the body. If silicone gel pads used to protect the pressure areas in theatre are inadequate by themselves to prevent occipital sores, surely they are even less likely to prevent injuries in the higher pressure areas of the pelvis and heels?

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Gebhardt KS, Bliss MR, Winwright PL, Thomas JM (1996) Pressure relieving supports in an ICU. *J Wound Care* **5:** 116–21

Bone marrow transplants in Crohn's disease

Sir,

Talbot et al (Vol 59(7), 1998, p. 580) described a patient with a 7-year history of steroid refractory Crohn's disease who went into long-term remission following an allogeneic bone marrow transplant for acute myeloid leukaemia. It might be anticipated that marrow transplants benefit patients with Crohn's disease as there is now strong evidence from animal and human studies to suggest that T lymphocytes play a key role in the underlying immune dysfunction associated with inflammatory bowel disease. It therefore seems plausible that replacement of aberrant mature lymphocytes would be advantageous to these patients.

Lymphocyte depletion has also been attempted in other ways. There have been conflicting reports in favour of lymphocyte apheresis in inducing and maintaining remission in active Crohn's disease (Bicks and Groshart, 1993; Lerebours et al, 1994) and ulcerative colitis (Sawada et al, 1996) and anti-CD4 monoclonal antibody given to patients with steroid refractory Crohn's disease appeared to improve the CDAI at 4 weeks (Stronkhurst et al, 1997). It has also been reported that Crohn's disease may improve in acquired immunodefiociency syndrome (AIDS) which is associated with loss of CD4+ lymphocytes (James, 1988; Popsai et al, 1994) although this finding is not consistent (Bernstien et al, 1994; Christ et al, 1994)).

In a report of uncontrolled observations, allogeneic bone marrow transplant for malignancy has induced long-term remission in 5 patients with Crohn's disease (Otero Bopez-Cubero et al, 1998). This seemingly beneficial response probably does not warrant the introduction of marrow transplantation as an alternative treatment as the risk of major complications is too high. Nevertheless these findings do serve to increase our own understanding of the pathogenesis of Crohn's disease, with focus on some of the early events in the inflammatory process.

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Correction

In the editorial 'Medical schools: their relationships with research institutions' (Vol 59(12), 1998, p. 912), an error occured within the text. The line that read 'In the UK, the RAE should continue to concentrate resources in those institutions that have already been successful with little prospect for the others to break out of the mould.', should have read 'In the UK, the RAE will continue...'.