Roberts *et al* describe difficulties in recruiting patients into peer-review teams. In our most recent cycle of 11 peer-review visits we were able to recruit a lay person (patient or carer) onto each reviewing team but only seven visits went ahead with lay involvement, due to unexpected ill health of the lay person or their family (data unpublished).

Roberts *et al* raised the issue of the cost-effectiveness of peer review. We would suggest that our regional approach over a longer period minimises the organisation required, and that a one-day visit every five years may be a relatively low cost exercise for staff for a clinical governance activity which may yield significant potential benefits including multidisciplinary education, revalidation, and improving patient care.

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Differential diagnosis of motor neurone disease

Editor – The review of motor neurone disease by Wood-Allum and Shaw (*Clin Med* June 2010 pp 252–8) was comprehensive and succinct. It did, however, miss the chance to highlight a very important differential diagnosis, Pompe's disease which is sometimes called glycogen storage disease type 2 or acid maltase deficiency. Recognition of this disorder is important because, although rare, it is now treatable with enzyme replacement therapy and the results are better the earlier treatment is initiated when muscle loss is minimal.

Pompe's is an autosomal recessive condition,[1] causing a deficiency of the lysosomal enzyme, acid maltase, (synonym, acid alpha-glucosidase) which degrades glycogen. This results in a build up of glycogen, particularly in skeletal muscle cells ending with cellular damage and destruction. When it occurs in juveniles or adults it presents with slowly progressive proximal muscular weakness and wasting, often involving the diaphragm and other respiratory muscles. Patients often languish for many years with an incorrect diagnosis such as benign progressive spinal muscular atrophy. The diagnosis should be entertained in all patients with slowly progressive proximal muscle wasting. Diagnosis is currently very easy, using enzyme assays on dried blood spots, leucocytes or cultured skin fibroblasts.[1]

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Reference

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Motor neurone disease: practical update ignores rehabilitative approaches – particularly assistive technology

Editor – The otherwise excellent review by Wood-Allum and Shaw (*Clin Med* June 2010, pp 252–8) made no mention of the role of the Motor Neurone Disease (MND) Association which supports patients, carers and professionals alike.[1] In some areas the MND Association has professional support workers who can work closely with the multidisciplinary team. This team may be supported by a consultant in rehabilitation medicine.

The authors recognised the importance of the multidisciplinary team but did not discuss the benefits such teams provide in ameliorating symptoms, particularly in the distressing later stages. Provision of assistive technology can have dramatic effects, eg electrically operated beds, wheelchairs (which can be powered) and environmental control units (ECUs).

Electrically-controlled beds facilitate: control of dependent oedema, transferring in/out of bed and management of limb pain which may/not be related to spasticity. In the later stages of MND the control knob may need to be fixed close to the patient's hand, or be operated by an ECU.[2] Carers benefit when patients' can themselves control body posture in bed and thus do not need to call for assistance when patient's want to change position. Not all beds can be operated by ECUs and specialist advice is needed to know which can.

Powered wheelchairs are valuable in overcoming problematic immobility for users[3–6] and assist their carers.[3,7] They have been available for indoor/outdoor use in the UK since 1996.[8] With sufficient technical support, these chairs can be controlled with integrated systems to enable communication, ECU and powered wheelchair to be controlled by a suitably sited control system.[9] EPIOCs can also have recline functions to facilitate swallowing and breathing from the optimal seated position. Additional tilt functions prevent sliding forward in the chair and facilitate pain and pressure management.[10] The mobile arm supports mentioned in the article can be fitted to either manual or powered wheelchairs.

ECUs have long been recognised as bringing benefits to those with progressive weakness which use about 20% of the ECUs provided in the UK. They facilitate independent operation of electrically operated devices, eg radios, heaters, lights, etc.

Assistive technology should be considered for those with progressive neurological weakness.

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Polymyalgia rheumatica

Editor – Dasgupta, writing on behalf of the polymyalgia rheumatica (PMR) guideline development group, presents a welcome and thorough overview of this common condition (*Clin Med* June 2010 pp 270–4). I have concerns with the recommended three-monthly 'lab monitoring' of full blood count, erythrocyte sedimentation rate/C-reactive protein (ESR/CRP), urea and electrolytes and glucose. The management of straightforward PMR is to relieve symptoms (and not to treat inflammation) until the condition runs its natural course. Steroid withdrawal should be based on the clinical picture and not on the level of ESR and this is alluded to in the article 'raised ESR/CRP without clinical symptoms is not an indication to continue corticosteroids'.

It is my belief, based on reviewing many patients with PMR and iatrogenic Cushing's/osteoporosis, that the main reason for the continuation of higher dose steroids is the regular checking of an ESR to follow disease activity. The secret to the successful management of straightforward PMR is, once the diagnosis has been made, never to check an ESR/CRP unless there is a clinical indication.

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In response

Editor – We thank Dr Morris for highlighting an important issue – the objectives of steroid treatment for polymyalgia rheumatica (PMR). Steroids are prescribed for their important effect on pain, disability and stiffness and the quality of life in untreated PMR is lower than in most other comparable conditions. On the other hand steroids also have many side effects and over-treatment based on raised inflammatory markers alone may prolong duration of treatment and induce treatment co-morbidities such as fractures, diabetes, hypertension, weight gain and cataracts.

However, we now know that the PMR constitutes only one of many conditions that can present with bilateral shoulder pain and stiffness. Such conditions include late onset rheumatoid arthritis, other arthropathies, spondyloarthropathies and connective tissue diseases. Large vessel vasculitis may also present with polymyalgia, constitutional symptoms and raised inflammatory markers. Other serious pathology, such as systemic infection, disseminated cancer and so on, may also be mistaken as PMR and may have an initial response to high dose steroids.

We therefore stand by our recommendation of inflammatory marker testing in the context of a clinical review – especially in the first year of disease. Transient elevations of CRP/ESR may be due to common causes such as urinary or chest infections and urinalysis and chest radiographs may be considered. Persistent elevation in the presence of definite symptoms suggests partial or non-response to steroids, search for alternative pathology or adjuvant immunosuppresives and a specialist referral. Persistent symptoms in the absence of elevated markers suggests evaluation of co-existing non-inflammatory conditions such as osteoarthritis, rotator cuff and other local shoulder conditions, fibromyalgia, etc. These conditions should be dealt with by explanation, reassurance and local treatments such physiotherapy, injections and exercises; while the steroid dose is tapered.

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A complicated hyperglycaemic emergency

Editor – I read with interest the article by Vidyarthi and Chowdhury describing a hyperosmolar non-ketotic diabetic emergency complicated by diabetes insipidus (*Clin Med June* 2010 pp 264–5).

I agree that these complex cases are best managed in a critical care environment where point of care testing is available to guide therapy. I feel a number of other features merit clarification, however. Firstly, the authors fail to emphasise that hyperglycaemia causes water shift from intracellular fluid (ICF) to extracellular fluid (ECF). Correction of hyperglycaemia thus causes in a influx of water back into the ICF causing a rise in serum sodium despite reduced free water losses. As this rise is accompanied by an influx of water into the brain, osmotic demyelination syndrome (central pontine myeliniolysis) should not arise, as long as serum osmolarity is falling. Conversely cerebral oedema can be a risk if serum osmolarity falls very rapidly with volume expansion. However, this danger may have been overemphasised in this case where serum osmolarity paradoxically rose with therapy, attributed by the authors to diabetes inspidus of uncertain aetiology. I feel administration of large volumes of 0.9% saline may have contributed to this outcome. The patient described was in early shock with a mean

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