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only seven visits went ahead with lay involvement, due to unexpected ill health of the lay person or their family (data unpublished).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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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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