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DRAMATIC DELAYED RECOVERY FOLLOWING SEVERE ANTI-NMDAR ENCEPHALITIS

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Introduction Long term outcomes in anti-N-methyl-D-aspartate receptor (NMDAR) encephalitis are reported to be favourable in 80% of cases, but there is little information regarding recovery following prolonged severe neurological disability.

Case 1 A 16-year-old female presented with headache, fever and acute encephalopathy. Anti-NMDAR antibodies were positive, with an ovarian teratoma. Treatment included oophorectomy, plasmapheresis, corticosteroids, cyclophosphamide, intravenous and intrathecal rituximab, and alemtuzumab. After 15 months hospitalisation, she remained in a vegetative state, with tracheostomy and percutaneous gastrostomy tubes, on multiple anticonvulsants. She received no further immunotherapy. MRI brain 2 years after illness onset demonstrated severe generalised cerebral atrophy. Three years after onset, she began to improve. At 5+ years she was seizure-free without anticonvulsants, fully independent and ambulatory, without overt cognitive deficits or personality change. MRI brain at 5 1/2 years demonstrated recovery of cerebral volume to within normal limits, but residual hippocampal atrophy.

Case 2 A 21-year-old female presented with headache, fever and personality change. NMDAR-antibodies were positive. A malignant ovarian teratoma was treated with salpingo-oophorectomy and chemotherapy (bleomycin/etoposide/cyclophosphamide). She received plasmapheresis, immunoglobulins, corticosteroids, intravenous and intrathecal rituximab, and bortezomib. She was finally discharged after 21 months hospitalisation with tracheostomy and gastrostomy tubes, severe behavioural disturbance, minimal functional limb movement, and fully dependent care needs. Clinical recovery began 2 1/2 years after disease onset, and at 4 years she is fully independent with residual cognitive deficits, engaging in volunteer work.

Conclusions These cases highlight the potential for dramatic delayed clinical improvement despite prolonged severe neurological disability in anti-NMDAR encephalitis.

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PRIMARY CUTIBACTERIUM ACNES CENTRAL NERVOUS SYSTEM INFECTION CAUSING FOCAL SEIZURES: AN UNUSUAL PRESENTATION OF A RARE DISEASE

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Introduction *Cutibacterium acnes* is a Gram positive, anaerobic bacterium of low pathogenic potential that forms part of the normal cutaneous flora. Although most often identified as a contaminant in culture of microbiological specimens, it is

commonly implicated in both postoperative wound and implantable device infection. Neurosurgical device infections secondary to *C. acnes* are well recognised and are likely secondary to bacterial contamination from the skin during surgery. Indolent infection characterised by delayed presentation of weeks to months following intervention is common. *C. acnes* infection involving the central nervous system (CNS) in the absence of previous neurosurgical intervention is rare, but has been described following dental or mastoid infections and following facial trauma. A further case series has reported de novo *C. acnes* CNS infection occurring in the absence of these recognised risk factors, but with clinical features of meningitis being common to all.

Methods and results We describe a unique case of primary *C. acnes* extra-dural collection in a previously well patient with no neurosurgical history presenting with sub-acute focal seizures and progressive focal leptomeningeal thickening on MRI.

Conclusion *C. acnes* CNS infection can occur in the immunocompetent and in the absence of neurosurgical intervention.

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A CRITICAL REVIEW OF BIOMARKERS FOR HEREDITARY SPASTIC PARAPLEGIA

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Introduction Hereditary spastic paraplegia (HSP) is a rare neurodegenerative condition characterised by lower limb weakness and spasticity. Currently, treatment is symptomatic and there is no disease modifying therapy. Though candidate therapeutic agents have been identified, sensitive biomarkers to measure treatment efficacy in clinical drug trials are lacking. There are many challenges in the search for appropriate biomarkers including the rarity of HSP, clinical and genetic heterogeneity of HSP and slow disease progression.

Methods We performed a search on PubMed and Medline using the search terms ('hereditary spastic paraplegia' OR 'spastic paraparesis') AND 'biomarker*'. We searched the reference lists of relevant articles to identify further studies. We collected data on number of participants, HSP genotype, methodology, and outcomes.

Results 72 papers were identified: 2 on Rating scales, 9 on gait analysis, 33 on neurophysiological measures, 23 on neuroimaging markers and 5 on biochemical markers. The studies reviewed demonstrated variation in methodologies and outcomes, including mixed genotype (41/72 papers) and genotype-specific (31/72 papers) patient cohorts, varied neurophysiological techniques and different outcome measures. 68/72 studies reviewed had small patient numbers (<50 patients). All potential biomarkers reviewed were able to differentiate HSP patients from controls. Only diffusion tensor imaging (DTI) parameters showed significant correlation with disease severity.