Preliminary report on corticosteroid therapy for ataxia-telangiectasia

Severe neurodegeneration is one of several clinical features of ataxia-telangiectasia (AT), yet control of CNS symptoms in patients with AT has received little attention. Buoni *et al.* have presented a case report in which corticosteroid therapy markedly improved the neurological symptoms of a 3-year-old boy with classic symptoms and molecular evidence of AT.

The boy was given the corticosteroid betamethasone at 0.05 mg/kg body weight every 12 h. The child's parents had previously noted improvement in neurological symptoms on the occasions when the boy had been treated with betamethasone for asthmatic bronchitis attacks. Here, improvements in neurological symptoms were evident after 2–3 days; after 4 weeks of treatment they were dramatic. The most notable adverse effects were increased appetite and body weight, and moon face. Two videos (available to subscribers at http://www.archneurol.com) illustrate the improvement in symptoms and the adverse effects over the 4-week period. After this time, methylprednisolone (1 mg/kg/12 h) was substituted for betamethosone, with no observed beneficial effect. The change in therapeutic agent was an attempt to provide long-term therapy, as the relatively long half-life of betamethasone precluded its use for this purpose.

The authors suggest a range of mechanisms by which corticosteroids could affect neurological symptoms, including regulation of corticosteroid-responsive genes and suppression of inflammation. They note that the use of these drugs in patients with AT, who are already immunosuppressed, requires careful consideration, and call for further work to investigate the cellular pathways of corticosteroid action in the CNS and the most appropriate treatment regimens for AT.

Original article Buoni S *et al.* (2006) Betamethasone and improvement of neurological symptoms in ataxia-telangiectasia. *Arch Neurol* **63:** 1479–1482

Electronic neural implant aids functional reorganization of the motor cortex

Therapies that stimulate the functional reorganization of the nervous system can benefit patients recovering from neural injuries and other movement disorders. Jackson *et al.* report on a study in which two macaque monkeys were fitted with autonomously operating electrical implants—termed 'Neurochips'. The implants connected two sites in the wrist area of the primary motor cortex and resulted in stable reorganization of motor output.

The Neurochip recorded action potentials at one electrode (Nrec), and delivered identical electrical stimuli at another (Nstim), thereby artificially connecting two distinct sites. After one or more days of conditioning through continuous operation under normal behavior, the response (measured as angle of wrist torque on intracortical microstimulation) elicited from Nstim was the same as that seen before conditioning. The response elicited from Nrec changed ($P = 0.0005$) to resemble that obtained from Nstim; the response from a control neuron showed no change. These findings indicated that a functional neural connection had formed between Nrec and Nstim; changes were consistent with the potentiation of synaptic connections between these artificially linked populations of neurons, and they persisted in some cases for over 1 week.

The authors conclude that artificial connections such as those provided by their Neurochip could act as neural prostheses, replacing and strengthening damaged pathways in cases of neural injury. Electronic devices could permit stable *in vivo* reorganization of cortical maps during normal behavior and contribute to the long-lasting rehabilitation of selected neural pathways.

Original article Jackson A *et al.* (2006) Long-term motor cortex plasticity induced by an electronic neural implant. *Nature* **444:** 56–60

Self-regulation of slow negative brain potentials can improve symptoms of ADHD

Stimulant medications for attention deficit hyperactivity disorder (ADHD) can cause adverse effects, have no proven long-term efficacy, and fail to be effective in about 25% of the pediatric conditions associated with the disorder. Children with ADHD are believed to have impaired ability to regulate their cortical excitation thresholds, displaying slowing of electroencephalogram oscillations in prefrontal regions and longer latencies as well as smaller amplitudes of