

## **ACQUIRED NEUROMYOTONIA/ ISAAC'S SYNDROME**

### **Acquired Neuromyotonia/Isaac's Syndrome**

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#### *Definition:*

Acquired Neuromyotonia is an immune disorder that presents with cramps and fasciculations (twitching or flickering of muscles) due to peripheral nerve hyperexcitability. The condition can affect all limb and trunk muscles. Speech can be involved and results in intermittent speech difficulties. The autonomic nervous system can also be affected and this may present as gastrointestinal symptoms with bloating, burping and altered bowel habits or excessive sweating. Patients may also complain of fatigue.

#### *Aetiology:*

The cause of acquired neuromyotonia is not known. An antibody to voltage gated K channels can be found in the serum of most individuals with the condition.

#### *Clinical Examination:*

Patients have normal muscle bulk and tone. Muscle power is normal. Cramps may be easily elicited with voluntary activity. Fasciculation of a variety of muscles may be seen and these may flit from one muscle to another.

#### *Diagnosis:*

The diagnosis is one of exclusion.

- 1) Neurophysiological tests are useful as they show fasciculation potentials with after discharges. Despite the fasciculations, needle tests of the muscle (EMG) do not show any other signs of denervation in the muscles. Myotonic discharges (distinctive bursts of muscle activity) can be seen on needle EMG. Sensory and motor nerve conduction tests are normal.
- 2) Voltage gated K channel antibodies may be present in the patient's blood.

#### *Treatment:*

Most patients respond well to treatment but treatment is usually needed indefinitely.

- 1) Ivlg: Intravenous immunoglobulin is made from pooled human serum or can be made in the laboratory. This contains many different proteins that are thought to bind with disease causing antibodies and relieve the symptoms. Patients receive intravenous infusions of the Ivlg at 4 weekly intervals.
- 2) Plasma Exchange: As this is more invasive and costly treatment, plasma exchange is reserved as second line treatment. This involves removing the patient's blood, washing it and returning the blood cells with added fluid to the patient (similar to dialysis machines for kidney disease).
- 3) Symptomatic treatment for the cramps is given in the form of anti-epileptic drugs such as carbamazepine.

#### *Prognosis:*

Occasionally patients go in to a spontaneous remission but most patients require ongoing treatment and are able to continue a normal life.