

In order to devise strategies to mitigate this acquired disability, a precise and quantitative description of the post-operative neurocognitive phenotype is necessary. This study is designed to assess the feasibility of using the KINARM robot to quantify the changes in the neurological function after cardiac surgery. *Methods:* Patients without prior history of cognitive dysfunction were recruited from the pre-operative cardiac surgery clinic, and underwent pre-operative assessment with the KINARM. The KINARM provides a quantitative assessment of the neurocognitive control of the upper limbs. During bypass surgery, brain tissue oxygen levels were measured with near-infrared spectroscopy. Patients were reassessed with the KINARM post-operatively at 3 months. *Results:* To date, 12 participants have been recruited (mean age = 65 years, all male). On straightforward tasks, such as visually guided reaching, the majority of patients scored within the normal range, both pre- and post-operatively. In more complex tasks, required visuospatial and executive functioning, post-operative deficits were more pronounced. *Conclusions:* It is feasible to use the KINARM robot to provide a quantitative measurement of the neurocognitive phenotype of patients after cardiac surgery.

NEUROMUSCULAR

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Bilateral facial nerve gadolinium enhancement and GBS

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Background: Facial diplegia with parasthesia (FDP) is a rare variant of Guillain–Barre syndrome (GBS), and has been reported in less than 1% of GBS cohorts. Here we describe a case of FDP with novel imaging findings and discuss the differential diagnosis. *Methods:* Case: A 39-year-old man referred to the emergency department with a 2 week history of right facial palsy progressed to bilateral facial palsy. His exam demonstrated severe, complete facial diplegia with only very mild limb weakness and present but diminished deep tendon reflexes. *Results:* Cerebrospinal fluid analysis showed albuminocytologic dissociation. Electromyography was consistent with a demyelinating process. MRI with contrast revealed bilateral enhancement of the facial nerves in the intracanalicular portion and in the region of the geniculate ganglion. A diagnosis of GBS was made and the patient was treated with IVIG. Over the course of several weeks the patient improved. *Conclusions:* Although nerve root enhancement of the spinal cord is described with GBS, nerve root enhancement effecting cranial nerves has only rarely been described. In addition, the relative limb-sparing with complete facial paralysis in this case is also an unusual phenotype. The Gadolinium enhancement of the bilateral facial nerves is thought to represent blood brain barrier breakdown due to GBS.

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Survey of Canadian myotonic dystrophy patients' access to computer technology

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Background: Myotonic dystrophy is an autosomal dominant condition affecting distal hand strength, energy and cognition. There is a neuromuscular patient portal under development that has the potential to give voices and resource access to patients, regardless of location via the internet and social media. The current state of access to technology and underlying factors affecting use and interest were explored. *Methods:* Surveys were mailed to 156 participants with myotonic dystrophy type 1 (DM1) through the Canadian Neuromuscular Disease Registry. The survey questions touched on demographics, technology use, internet use, and interest in the portal. *Results:* Seventy-two participants (43 female) responded so far and 50% were younger than 46 years. Most (62/72) used the internet and 94% of participants had access to an internet-connected device. Almost half of the responders (34/72) used social media. The complexity and cost of technology were commonly cited reasons not to use technology. The majority of responders (79%) were interested in a myotonic dystrophy patient portal. *Conclusions:* DM1 patients have access to and use technology such as computers and mobile phones. They expressed interest in a portal that would provide them with access to relevant information such as guidelines, self-management modules, educational videos, and support groups.

P.057

Distal hereditary motor neuropathy type I due to the GARS: c.1415A>G, p.His472Arg mutation

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Background: The distal hereditary motor neuropathies (dHMN) have been characterized through case reports and family studies. Their genetic characterization remains a work in progress. An appreciation of the genetic underpinnings may lead to treatment options in the future. Hence reports, like this one, which add to this understanding, remain extremely important. *Methods:* The clinical presentations, electrophysiology and genetics of two patients with the dHMN I phenotype are described. *Results:* A mother and son presented with slowly progressive distal weakness of the lower extremities with onset in the first and second decades. Distal weakness of the upper extremities developed later. Examination 20 and 50 years after onset revealed wasting and weakness of distal upper and lower extremity muscles with absent distal and preserved proximal deep tendon reflexes. Sensory examination was normal. Electrodiagnostic studies demonstrated a motor axonopathy. Genetics testing revealed a missense mutation on chromosome 7, exon 11 of the GARS gene: c.1415A>G (p.His472Arg). *Conclusions:* GARS mutations have been identified in patients with the dHMN I (juvenile onset distal weakness and wasting) and dHMN V (upper limb predominant) phenotypes. However, this mutation has not previously been directly linked to the dHMN I phenotype.