

CPSP launches study of ‘moving target’ disorder

Along with the substantial physical and emotional toll of any chronic disease, patients with paediatric myasthenia and their families have to endure the general lack of awareness of the condition.

The Canadian Paediatric Surveillance Program (CPSP) welcomed the new year with a new study that aims to help address this.

Dr. Hanna Kolski, Director of the Glenrose Pediatric Neuromuscular Clinic and Associate Professor at the University of Alberta, is the study’s principal investigator. While study findings aim to inform best practices, she says, “paediatricians can begin making a significant impact on the quality of life for those with the condition right away. The key is to identify it early.” Those who have been able to do so are already managing the condition with their patients.

Paediatric myasthenia is very hard to diagnose, Dr. Kolski continues, because the symptoms may fluctuate from minute to minute and day to day. “I’ve called it a ‘moving target.’ The presentations of this disorder are off the beaten track.”

As a result, she says, “these are the kids in gym class that are pushed, and can’t keep up.” Yet she and her colleagues see those children months or years after they have symptoms. And since about half the cases she sees are ocular alone, they are mostly referred by ophthalmologists.

Early interventions yield striking improvements

If unrecognized and advanced, paediatric myasthenia can become more serious, possibly leading to respiratory weakness and, in rare instances, death.

However, if recognized and treated early, the prognosis is quite positive. Even more encouraging, Dr. Kolski notes that most children will improve soon after they start with treatment, and about one-third will be cured. “But none of that can happen until the index of suspicion is raised.”

“My goal is to spark a potential diagnosis,” she says. “If it’s a clear-cut case where they see ptosis, double vision and high antibodies against acetylcholine receptors, physicians should be tuned into it. If there is limb and body weakness, paediatricians can dig a bit deeper into the *pattern* of the fatigue, along with the potential change in symptoms day and night.”

In this quite characteristic or classical presentation, she adds that it would be “very reasonable” for a paediatrician to

Is it paediatric myasthenia?

Main symptoms may include:

- rapid fatigue
- muscle weakness (worse with repetitive activities)
- ptosis of the eyelid
- double vision
- trouble swallowing during feeding
- respiratory difficulties
- poor head control in infants
- delayed development

treat the child. But if what they see is atypical, they should refer the patient to a paediatric neurologist.

The study will collect data through the CPSP, and also collaborate with paediatric neurologists across the country. A progress update is expected in one year’s time.

You can view the study protocol on the CPS website: <http://www.cps.ca/English/surveillance/cpsp/studies/PM.pdf>. 📄

Olympic moment for CPS member

Dr. Aline Levi, a CPS member and family physician in Dollard-des-Ormeaux, Que., carried the Olympic Torch on December 6, 2009 along its relay route through Granby, Que. It was a fitting role for this mother of five, who also swims competitively and works as a paediatric rehabilitation consultant in Laval.

The Torch made a 106-day trek across Canada before reaching the opening ceremonies of the Vancouver 2010 Olympic Winter Games at BC Place Stadium on February 12.

