New pediatric heart failure guidelines a first in Canada

October 30 2012

The Canadian Cardiovascular Society is the first in Canada to issue guidelines aimed at helping primary care and emergency physicians, as well as specialists, recognize and manage heart failure in children. The guidelines were released today at the Canadian Cardiovascular Congress.

Pediatric heart failure is often fatal and occurs in about 3,000 children annually in North America. Worldwide, the problem is far greater and the causes are diverse. To date there has been little guidance to assist practitioners who deal with children with heart failure.

"The previous guidelines, produced in 2004 by the international Society for Heart and Lung Transplantation, are now dated and were not designed for front-line practitioners who deal with these children at the first encounter," says Dr. Paul F. Kantor, who chaired the guidelines and is head of pediatric cardiology at the Stollery Children's Hospital, University of Alberta.

"Providing guidance in this area may help to solve one of the biggest challenges we have: that children with heart failure are usually not recognized early and treated effectively. When heart failure presents late in the disease course, it is more dangerous and can be fatal."

Heart failure in children is far more likely to cause death than cancer, but the problem is not nearly as well recognized, says Dr. Kantor.

About half of the children who present with obvious heart failure will

die, or require a heart transplant within five years. Unfortunately, awareness regarding organ donation is still relatively low among Canadian doctors and patients, and a heart transplant is not always available.

"We try our best to treat them with medication and we also use advanced devices such as the Berlin Heart and other ventricular assist devices to keep them alive. Occasionally recovery occurs and some patients will be fortunate enough to receive a transplant," Dr. Kantor says.

"The biggest issue we face is that our patients are often presenting very late and with advanced disease. The earlier they are diagnosed, the more likely it is that we will be able to offer effective treatment."

The new guidelines will provide a framework for early recognition and treatment.

The key recommendations for early recognition are:

- Cardiomyopathy, or heart muscle disease, is one of the main causes of heart failure in children and should be excluded when a child presents with unexplained rapid heart rate or rapid breathing. This may be a familial condition caused by one of several gene abnormalities.
- Myocarditis, a viral infection of the heart muscle, may be present when children present with abdominal pain and vomiting and have signs of poor circulation.
- Specialized pediatric echocardiography is required to exclude heart failure and should be obtained in children with unexplained symptoms and signs of abnormal circulation.

"Often children are brought to the emergency room with shortness of breath and some cough and are thought to have asthma, when in fact

they have very severe heart failure," says Dr. Kantor. "The clues of a very unusually fast heart rate and low blood pressure are sometimes overlooked and these children will be sent home with a 'puffer' for their breathing problems, which are actually due to heart failure."

In the same way, he says, teenagers who come to the emergency room with nausea and vomiting and abdominal pain may be thought to have gastroenteritis but may actually have myocarditis. "This is one of the earlier considerations that people in the emergency department should make, since the earlier this is diagnosed, the better the outcome."

Pediatric echocardiography is a specialty in its own right and is different from adult echocardiography. Unfortunately, echocardiography is sometimes done in a non-expert setting, which can lead to misdiagnosis and delay in starting the correct treatment, Dr. Kantor emphasized.

Key recommendations for management after diagnosis are:

- Admission to a hospital that has pediatric expertise for immediate care.
- Ongoing care to be provided by an expert in pediatric cardiology.

"It is very important that these children receive expert care because they tend to deteriorate rapidly," says Dr. Kantor. "They need to be admitted to a hospital that has pediatric expertise and be evaluated by a children's heart specialist, with echocardiography done by that specialist."

Key recommendations for treatment are:

- Prompt use of diuretics.
- Treatment with inotropic drugs to rescue the patient, and a transition to ACE inhibitors thereafter.

"We strongly recommend the use of diuretics, which are very effective in an emergency setting," says Dr. Kantor. "We also strongly recommend rescue treatment with inotropic drugs, such as epinephrine and milrinone, to restore the circulation, followed later on by ACE inhibitors, which are effective heart failure treatment in adults and appear to be effective in children as well."

For myocarditis, the experts recommend supportive care, to allow the heart to recover on its own. This can mean giving the patient drugs to support blood pressure, circulation, and occasionally, use of a ventricular assist device, to improve the circulation while the heart recovers, giving it a chance to rest.

These guidelines will benefit practitioners in the field by standardizing practice across institutions, and allowing patients to benefit regardless of where they are cared for.

"Canadian Cardiovascular Society guidelines are an invaluable resource that establish best practices in patient care," says Dr. Michelle Graham, chair of the CCS Guidelines Committee. "These guidelines are important references for our healthcare practitioners in Canada and they are recognized and used by practitioners around the world."

A family's story Tim and Theresa Miller from Delaware, Ont., lost their teenage son to heart failure in 2003.

"We got Daniel to hospital twice on a Monday, and then on the Wednesday. He had all the classic signs that they talk about in these guidelines and they were not recognized as heart failure," says Tim Miller. "Finally, they were trying to resuscitate him and the doctor said, 'It doesn't look good. He probably has myocarditis.' But by then it was too late. He had been in hospital for almost two full days."

"He was 17," adds Theresa Miller. "He was a normal, healthy, bright kid. He'd just finished his application for university, and then, just out of the blue, he got this case of the flu, but he was sicker than we had ever seen him. It was different from any other flu he'd ever had."

"It's the regular physician on the floor that needs to be aware of these guidelines to get the patient on the right track," adds Tim. "The guidelines talk about unusually high heart rate, and that was one of Daniel's symptoms. When he went in that first day, the doc came back and said, 'Wow, his heart rate is really high.'"

"And it was at that moment in time, if these guidelines were in place and had sparked that doctor to recognize the symptoms of myocarditis, that was Daniel's chance. And he was at a tertiary care facility, all the experts were there, but they looked at him as someone having the flu and never thought to look at him broadly enough and treat him in the way that he needed to be treated," says Theresa. "We think these guidelines could be hugely helpful because they give logical steps of what needs to happen if we are going to give these kids a chance."

"This was totally out of the blue, that on a Saturday you have a son, and by Thursday, you don't."

Provided by Heart and Stroke Foundation of Canada