Canadian researchers bring new insights into Kawasaki disease and its consequences

(January 2017) — Kawasaki disease (KD) is the leading cause of acquired heart disease in Canadian children. While some of the approximately 500 children diagnosed annually in Canada will have no lingering effects, some will develop lifelong heart problems. Recent publications by researchers located, primarily, at The Hospital for Sick Children in Toronto (SickKids) and at Centre Hospitalier Universitaire Ste. Justine (CHU-SJ) in Montreal, are beginning to peel back some of the mysteries surrounding Kawasaki disease, helping both patients and their families.

A recent publication by nurse practitioner Nita Chahal (SickKids) and her colleagues explores some of the psychosocial factors impacting the lives of children and their families when Kawasaki disease results in coronary artery aneurysms (CAA). CAAs result in many intrusive procedures, require constant management by children and their parents, and can leave children and parents feeling uncertain about the future. While this results in a considerable burden of struggle for families, Chahal et al. also found a “cautious optimism,” which increased where families had consistent and reliable information about their child’s health.

Newly published research by SickKids’ Dr. Brian McCrindle and colleagues re-affirms the importance of early diagnosis. If treated within ten days of the onset of Kawasaki disease, intravenous immunoglobulin can reduce the chances of developing CAA. The research found, however, other factors that can affect the outcome.

Dr. Rae Yeung, also at SickKids, identified regulation of calcium mobilization as fundamental to the underlying biology of Kawasaki disease leading to a unique IL-1β signature, totally changing the way we think and treat Kawasaki disease as we already have powerful medications to block IL-1β. Her group identified a novel function for a gene, ITPKC, that controls calcium mobilization but more importantly controls the response to IVIG treatment. Along the same theme, Dr. Yeung together with a group of international colleagues, examined genomic data from children with Kawasaki disease from around the world, including North America, Europe and Japan, identifying a form of another gene responsible for the regulation of calcium ion levels through channels in the cell membrane, SLC8A1, that is associated with both Kawasaki disease and CAAs.

In Montreal, a team led by Dr. Nagib Dahdah and located principally at CHU-Ste-Justine examines important clinical questions in patient populations in Canada based on novel insights from the Kawasaki disease community more broadly. They have shown, for example, that the presence of a form of natriuretic peptide (an inflammatory marker) can add to the diagnostic criteria set out by the American Heart Association for KD and can be especially helpful in the diagnosis of incomplete KD. In the case of KD patients with CAAs, Dahdah and his team found that coronary artery bypass grafting (CABG)—often multivessel--was preferable to percutaneous coronary intervention (PCI).

Drs. Dahdah, Chahal, McCrindle and Yeung have also been authors of several other papers on Kawasaki disease published in the past year. See the list of publications below, representing just the past year.

Kawasaki Disease Canada, a registered charity, focuses on increasing awareness to enable early diagnosis, supporting families affected by KD, and supporting new research. Today, Kawasaki Disease Canada announces its second Student Research Award, with a deadline for applications of February 22. The Inaugural Student Research Award winner was Mathew Mathew, also at SickKids. Mathew's research involves constructing models of the hearts of KD patients with large coronary aneurysms and using computers to see how the blood flows through them. This is expected to help determine how the location and shape of the aneurysm affects the chances of forming blood clots.

Early diagnosis is critical in treating Kawasaki disease but the symptoms aren't always straightforward. Typical Kawasaki disease presents as a prolonged fever lasting five days or more with at least four other symptoms, including:

• a rash

• bloodshot eyes

• bright red, swollen, cracked lips

• "strawberry tongue"

• swollen hands and feet

• redness of the palms and soles of the feet

• swollen lymph nodes in the neck

However, Kawasaki Disease Canada encourages parents to ask their doctor about Kawasaki disease when a prolonged fever is accompanied by any two of the above symptoms.

About Kawasaki Disease Canada

Kawasaki Disease Canada is a national non-profit and registered charity with a mission to promote awareness and education about Kawasaki disease, support individuals and family members affected by the disease, and advance research into the disease and its consequences. For more information, visit kdcanada.org.

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