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Editorial

Finding Kawasaki Disease

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See article by Manlhiot et al., pages 303–309 of this issue.

Epidemiologic research on Kawasaki disease (KD) is fraught with many hazards, not the least of which is the lack of a definitive diagnostic test. Thus, epidemiologists must rely on clinicians to recognize the classic features of the illness, gather supporting laboratory data, and eliminate other competing diagnoses as appropriate. This essentially guarantees that there will be over- as well as underdiagnosis of KD. Added to this problem, most countries or regions do not have prospective surveillance programs. However, all patients with acute KD must be hospitalized to receive the standard treatment of high-dose intravenous immunoglobulin (IVIG). Thus, case ascertainment can rely on hospital discharge records or administrative databases. On the brighter side, KD owns its International Classification of Diseases, 10th Revision code (M30.3) so at least if the coding is done correctly, it will link to the correct diagnosis. A potential pitfall in coding is that patients might be admitted to the hospital with the working diagnosis of presumed KD, but results of subsequent testing suggest an alternative diagnosis and the KD diagnosis is abandoned. This is at least 1 potential source of coding errors. The coding of outcomes of interest, such as coronary artery aneurysms, is less straightforward because definitions on the basis of various national guidelines from Japan, the United States, and the United Kingdom vary in terms of using absolute internal dimensions of the artery vs measurements normalized for body surface area and expressed as standard deviations from the mean (Z score).[1-3]

In this issue of the *Canadian Journal of Cardiology*, a Canadian group from research institutes in Ontario present an update on the epidemiologic patterns of KD across Canada using the Canadian Hospital Discharge Database and comparing the results with a questionnaire-based survey performed across Canada in 2006 and 2009.[4] Their epidemiologic investigation had 3 main goals: (1) to compare questionnaire-based surveillance results and administrative database analysis for tracking of KD cases in Canada; (2) to update incidence data for Canada through 2014; and (3) to analyze regional and seasonal differences in KD incidence. The authors report a minor overestimation of approximately 7% using the administrative database compared with the questionnaire-based surveillance and concluded that relying on administrative databases with manual curation was a useful and accurate tool to track KD. Cases eliminated by manual curation included interhospital transfers, acute readmissions, and repeat admissions for cardiac care.

In contrast to the rising KD incidence reported from Asia, the authors report a stable rate of KD in the youngest cohort with only slightly increasing incidence among older age groups. The overall incidence of 19.6 cases per 100,000 children younger than 5 years is similar to the incidence reported for the United States.[5] Seasonal and regional variation in incidence rates across Canada were described. Intriguingly, the provincial level data showed a marked variation in incidence with those provinces bordering the Pacific and Atlantic Oceans and the Great Lakes having the highest rates. Because the environmental trigger of KD remains a mystery, perhaps this is a clue worth noting. The current paradigm for KD is that children are born with a genetic pattern that determines susceptibility as well as coronary artery outcome. The disease becomes manifest when these susceptible children encounter the environmental trigger and the event is "immunizing" in that recurrences are rare (1.3% in the Canadian study). The seasonality of KD in Canada mirrors that of Japan and other countries across the Northern Hemisphere with the highest incidence in the winter months of January through March and a dramatic nadir in the late summer/fall (August through October).[6,7] It might lead to further insights into the environmental trigger of KD to compare the seasonality according to province to ascertain if the different regions are synchronized, suggesting a large-scale exposure across Canada, or unsynchronized, suggesting more regional or local exposures that vary over time. Synchronized exposure would be in keeping with a hypothesis regarding large scale wind patterns from Asia associated with the seasonal rise and fall of cases in Japan.[8]

With respect to cardiovascular outcomes, 3.5% of the study population were reported as having aneurysms. As discussed previously, variation in definitions of aneurysms might contribute to differences in coding and more uncertainty

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See page 237 for disclosure information.

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about the exact number of patients with coronary artery damage. Still, this overall figure mirrors the results of the original clinical trial of IVIG in 1986, which reported a similar rate of aneurysms in IVIG-treated patients.[9]

Overall, the results of this study provide reassurance that analysis of the Canadian Hospital Discharge database is an accurate way to track KD cases in Canada and monitor trends over time. The accessibility and quality of this database will prove to be a valuable resource to support future studies of KD epidemiology in Canada.

Disclosures

The author has no conflicts of interest to disclose.

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