



## Editorial

# The Canadian Journal of Cardiology: Open and Growing

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*Canadian Journal of Cardiology (CJC)* enters its 35th year of publication in 2019. Founded and first edited by Dr Robert Beamish in 1985, and subsequently stewarded by Eldon Smith from 1997 until I assumed the role of Editor-in-Chief in 2010, *CJC* has evolved into a premier international cardiology journal. With approximately 1800 submissions in 2018, *CJC* had a 2017/2018 impact factor of 4.524 (a 3.4-fold increase over the 2009 value of 1.324) in the top quarter of cardiology journals, and a 5-year impact factor of 4.035. In 2017, *CJC* had more than 800,000 full-text article downloads and a 7.3-day average time from submission to first decision: one of the best in the area.

### **CJC Open**

*CJC* has been a pioneer in a number of areas and continues to blaze new trails. One exciting new development is the launch of our sister Open Access journal, *CJC Open (CJCO)*. After an extensive search and careful review of a number of excellent candidates, the selection committee chose an outstanding person, Dr Michelle Graham from the University of Alberta, as the first Editor-in-Chief of *CJCO*. Michelle has been a consistent pioneer and trail blazer in cardiology. She was one of the first women to brave the then male-dominated area of cardiology and added “insult to injury” by excelling in the academic domain that, until then, very few women had chanced. Her contributions to cardiology in Canada and throughout the world have been exceptional. She has performed ground-breaking work in the areas of emergency cardiology, geriatric cardiovascular medicine, new models of care delivery, health care outcomes, and many others. Michelle has served the Canadian Cardiovascular Society (CCS) extensively, most notably as chair of the Guidelines Committee, for which she played a key role in organizing, structuring, and strengthening process and output. Before becoming *CJCO* Editor-in-Chief, Michelle Graham was an

outstanding Associate Editor for *CJC*. I will always cherish the memory of Michelle telling me that she felt the need for a new challenge when her mandate as CCS Guidelines Committee Chair ended and asking me if I could find something for her to do at *CJC*. I suggested to her the possibility of joining the editorial staff as an Associate Editor. That was best decision I ever made as Editor-in-Chief of *CJC*.

Under her stewardship, *CJCO* is flourishing. Since the launch of the *CJCO* manuscript-handling website on October 5, 2018, just more than 2 months ago, 27 papers have been submitted: 16 directly and 11 as transfers from *CJC*. Of these, 8 have been accepted, 5 have been rejected, 8 have received requests for revision, and 6 are in the active-review process. Of the 7 articles in the inaugural January 2019 issue, 4 are Original Research papers, 2 are Images in Cardiology articles, and 1 is a Case Report. *CJCO* is accepting submissions for the same article categories as *CJC*; full *CJCO* Author Guidelines are available at <https://www.elsevier.com/journals/cjc-open/2589-790x/guide-for-authors>. Certain types of papers may be particularly appropriate for *CJCO* and more difficult to get into *CJC* because of space limitations: for example, trial design articles, reports of health care outcomes of primarily local interest, viewpoint papers, and valuable but primarily confirmatory studies in basic or clinical science. Articles submitted to *CJC* that are found to be of potential value but cannot be accommodated in *CJC* because of insufficient page availability are offered transfer to *CJCO*. For situations in which the papers have already undergone primary review at *CJC*, if transfer to *CJCO* is accepted, the articles can be submitted directly as revisions to *CJCO* along with any necessary modifications and responses to the *CJC* reviews on the original submission.

As *CJCO* is an Open Access journal, the articles published in it are fully available to all potential readers without the need for a subscription or download fee. Granting agencies are increasingly requiring Open Access publication, a development that points toward Open Access publication as the way of the future.<sup>1</sup> Open Access articles require a publication payment to offset the loss of revenue from subscriptions and download fees that otherwise cover publication costs. *CJCO* publication fees are relatively modest, running from \$900 (\$700 for CCS members) for Case Reports, Images, and Viewpoint articles to \$2100 (\$1600 for CCS members) for

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full Original Articles, Study Design papers, Review articles, and Systematic Review/Meta-analysis articles (all figures are in US dollars, as the *CJCO* publisher is based in the United States). *CJCO* fees are approximately 20% lower than comparable area industry average for full papers and 50% lower for Case Reports/Images/Viewpoints. For CCS members, the fees are approximately 30% lower for full articles and 68% lower for Case Reports/Images/Viewpoints.

### Trainee Initiatives

*CJC* is the official journal of CCS. One major component of its mission is to contribute to the academic development of young cardiovascular clinical/basic scientists in Canada. Consequently, *CJC* has developed—in collaboration with CCS—a series of initiatives to promote the involvement in *CJC* publication of trainees in Canadian cardiovascular training programs. There are 3 primary components to this training initiative.

#### Trainee article publication

*CJC* has developed an article publication training program initiative, in which Canadian Cardiology Training Programs are invited to name up to 2 trainees who will submit invited papers to *CJC* with the assistance of a designated local academic mentor. The first cycle was completed in 2015, with 11 trainee papers published, and feedback was positive. A second cycle followed, in which 10 centres throughout Canada agreed to participate and submit a total of 18 papers between June 2016, and March 2017. Of these, 15 papers were received (9 of which were accepted; 2 were rejected, and 4 are still in revision). The first 2 cycles involved cardiology programs only; a third cycle is currently under way, involving cardiac surgery and pediatric cardiology programs in addition to cardiology. For the current cycle, 13 centres are participating, with 36 articles projected; 15 have already been submitted, and 21 more are to be submitted through the spring of 2019.

#### Trainee editorial board members and trainee reviewers

*CJC* has initiated a program to include Canadian cardiovascular trainees as trainee members of the *CJC* Editorial Board or reviewer database. Each trainee has a mentor—in most cases, a member in good standing of the *CJC* editorial board—to support the trainee in article review and help to ensure quality control. We received 25 excellent applications in response to a call in 2016. After thorough grading by a selection committee, 7 trainees were selected as Trainee Editorial Board Members and 18 were added as trainee members of the *CJC* reviewer database. Their mandates began in January 2017 and ended in December 2018. The 7 trainee editorial board members reviewed an annual median of 12 papers (range 6 to 22) with a median time from review assignment to completion of 6 days (range of average times 0 to 12 days). The trainee reviewers reviewed a median of 5 papers per year (range 0 to 21) with a median turnover time of 7.5 days (range 2 to 13). Feedback from reviewers and mentors about the interest of the program and its value to trainees was positive. The quality of trainee reviews was evaluated by the *CJC* editorial staff as varying from “very good” to “outstanding.” A new set of 7 trainee editorial board

members was selected in the fall of 2018, including—for the first time—2 CCS members-in-training engaged in overseas cardiology fellowships, along with 22 trainee reviewers; they will begin their 2-year terms in January 2019, and we look forward to continued success of this innovative program.

#### Trainee section

CCS and *CJC* have initiated a Trainee Section that is run by trainees, with articles of particular interest for trainees written by trainees that deal with such matters as challenges in training, helpful advice for trainees, and trainee-oriented discussions. The Trainee Section appears in the CCS Society Pages of the journal and is managed by a committee consisting of 2 trainee co-editors (Jacinthe Leclerc and Erin Rayner-Hartley) and 2 faculty supervisors (Paul Poirier and Parvathy Nair). Seven articles have appeared in this section to date.

#### Focus on Congenital Heart Disease and Pediatric Cardiology

*CJC* has developed a special interest in congenital heart disease (CHD) and pediatric cardiology. These are subjects in particularly rapid growth and are, in our opinion, somewhat underserved in the current cardiology literature. Dr Kevin Harris, of the University of British Columbia, has been identified as a particularly valuable resource person in this area and has been recruited as an Associate Editor for *CJC*. Between June 2018, and now (December 2018), *CJC* has published 38 articles dealing with CHD and/or pediatric heart disease as the primary focus, constituting 17.6% of all *CJC* papers in this period. Space does not permit a full summary of these papers here, but we will briefly mention the content of selected articles to give a sense of some of the areas touched upon.

CHD is the most common birth defect, complicating about 1% of live births.<sup>2</sup> Islam et al used the Alberta Pregnancy-Birth Cohort to perform a population-based analysis of health care utilization among almost 450,000 children born in Alberta between 2005 and 2014, including almost 7000 with CHD.<sup>3</sup> The rate of hospitalization in the first year of life (excluding birth hospitalizations) averaged 235 per 100 children with single-ventricle physiology, 100 in moderate to complex CHD, 52 in simple CHD, and 9 in children without CHD. Outpatient visits during the first year followed a similar trend, with 4871 per 100 children with single-ventricle, 2278 for moderate to complex, and 1416 with simple CHD vs 246 per 100 children without CHD. The health care resource implications of these findings are clearly enormous, requiring strategies to optimize long-term care and resource planning for the CHD population and in the context of rapidly advancing health care technologies.<sup>2</sup> Beland et al review the history of CHD nomenclature in Canada and propose the adoption of a diagnostic list put forward by the International Society for Nomenclature of Paediatric and Congenital Heart Disease.<sup>4</sup> They also provide French-equivalent terms for all diagnoses, important for national research programs in a bilingual country such as Canada.

Arslani et al review the time course and occurrence of cardiac complications of CHD (including atrial fibrillation [AF] and/or flutter, ventricular tachycardia, atrioventricular

[AV] block, heart failure, endocarditis, stroke, myocardial ischemia/infarction, and pulmonary hypertension) among 2731 patients with CHD.<sup>5</sup> Just under one third (28%) had at least 1 complication. More than two thirds of complications occurred in adult life (> 18 years old); however, certain complications—such as perioperative stroke and complete AV block—were more common in childhood. Atrial tachyarrhythmias are an important complication of CHD, with increasing rates of AF after age 50. In the *CJC* Theme Issue on AF, Ebrahim et al provide a contemporary review of the mechanisms, epidemiology, and treatment aspects of AF in patients with CHD.<sup>6</sup> Reiner et al analyze the early precursors of an increasingly recognized complication of CHD: atherosclerotic heart disease.<sup>7</sup> They find increased carotid intimal-medial thickness in children with CHD vs age-matched controls, with the largest values associated with aortic coarctation and congenital transposition of the great arteries with arterial switch procedures.

A series of articles deal with congenital and pediatric coronary artery abnormalities. Kukuchi et al describe a case in which accelerated atherosclerosis caused by a congenital myocardial bridge led to myocardial infarction.<sup>8</sup> Isorni et al describe the unusual coexistence of 3 different types of congenital coronary abnormality in a single patient.<sup>9</sup> Kawasaki disease is an inflammatory vasculitis occurring in children, with coronary artery involvement in approximately 25% of patients not given anti-inflammatory therapy vs approximately 4% of treated patients.<sup>10</sup> Coronary artery aneurysms are a common complication. Dionne et al describe the use of optical coherence tomography to study the underlying pathophysiology of coronary artery involvement, with fibrosis, cellular infiltration, and intimal hyperplasia being ubiquitous findings and calcification, medial necrosis, neovascularization, and thrombus formation strongly associated with the development of aneurysms.<sup>11</sup>

Familial hypercholesterolemia (FH) due to mutations in lipid-metabolism proteins is the most common cause of premature atherosclerosis. To be optimally effective, treatment should begin in childhood, around age 10.<sup>12</sup> Perhaps because of the complexity and difficult accessibility of some of the criteria needed to diagnose FH according to standard criteria, many cases go unrecognized until it is too late. Ruel et al propose a simplified diagnostic definition for FH based on the distribution of low-density lipoprotein cholesterol levels.<sup>13</sup> The definition is adapted to the Canadian medical context and has the potential to lead to earlier diagnosis and therapy, a critical concern.<sup>12</sup> Brunham et al provide an updated CCS Position Statement on FH.<sup>14</sup> Among other aspects, they provide guidance for early diagnosis; the use of statins, ezetimibe and PCSK9 inhibitors; and improved health care delivery by translation of knowledge, mobilization of patient sensitization/support, and optimal provision of resources. They emphasize the fact that the risk of atherosclerotic heart disease is increased 10- to 20-fold by FH, whereas initiation of treatment in childhood or in young adults can normalize life expectancy. Rzek et al review the efficacy of PCSK9 inhibitors in FH, highlighting the importance of their optimal use if needed to obtain good control of cholesterol in this high-risk population.<sup>15</sup>

Several studies provided novel insights into the genetic basis of CHD. Iacocca et al describe a familial FH syndrome

involving extremely high LDL cholesterol and PCSK9 levels and severe resistance to statin therapy due to a duplication in the PCSK9 gene.<sup>16</sup> This finding has important implications for the understanding of FH and for the management of affected persons.<sup>17</sup>

Greenway et al describe a therapeutic trial of digoxin, showing striking efficacy for improving cardiac function in patients with a rare form of mitochondrial cardiomyopathy, the dilated cardiomyopathy with ataxia (DCMA) syndrome caused by mutations in *DNAJC19*, which encodes a protein that is part of a complex implicated in ATP-dependent transport of chaperone proteins from the inner-cell membrane to the mitochondrial matrix.<sup>18</sup> In an accompanying editorial, Song et al underline the therapeutic and conceptual importance of these findings and the potential to lead to advances in our understanding of the pathophysiology of mitochondrial cardiomyopathies.<sup>19</sup> The co-occurrence of cardiomyopathy with neurofibromatosis is rare. Kizawa et al describe a 2-year-old child with genetically based neurofibromatosis and restrictive cardiomyopathy, who proved to have an independent mutation in a titin gene causing the cardiomyopathy.<sup>20</sup>

A number of studies deal with novel diagnostic and therapeutic approaches. Hazari et al compare echocardiography and cardiac magnetic resonance (CMR) imaging in the longitudinal evaluation of myocardial mass in patients with Fabry disease.<sup>21</sup> They note that left ventricular mass, as measured by CMR, increases with the development of cardiac fibrosis and that enzyme-replacement therapy slows these changes, pointing to the value of this indicator. Kato et al apply automated, real-time 3-dimensional volume colour-flow Doppler echocardiography to obtain single-beat right-heart flow measurements in children with atrial shunts.<sup>22</sup> The feasibility of single-beat right-sided stroke-volume measurements provides a potentially valuable noninvasive index for children with CHD. Kowalik et al evaluate the use of several candidate biomarkers in predicting outcomes in adults (mean age: 36) with congenitally corrected transposition of the great arteries, concluding that a combination of high-sensitivity troponin values and systemic right-ventricular end-diastolic areas is the best predictor of adverse cardiovascular outcomes, with an area under the receiver-operating curve (AUC) of 0.79.<sup>23</sup> Mocerri et al describe changes in right-ventricular function during pregnancy in repaired tetralogy of Fallot, indicating the potential value in counselling and follow-up.<sup>24</sup> The Ross procedure, involving the use of pulmonary valve autografts in the aortic valve position in conjunction with pulmonary valve bioprosthesis placement, is increasingly being used in the management of aortic stenosis, particularly because of congenital bicuspid valves in young adults.<sup>25</sup> Kellermair et al describe the case of 51-year-old man who had undergone a Ross procedure for congenital aortic stenosis and presented with severe right ventricle-to-pulmonary artery conduit stenosis/regurgitation. A Melody-system pulmonic valve (Melody Transcatheter Pulmonary Valve, Medtronic, Fridley, Minnesota) was implanted via a transcatheter approach; the patient developed severe signs of right-ventricular outlet obstruction due to prosthetic-valve thrombosis within 24 hours, which ultimately required surgical valve replacement.<sup>26</sup> The thrombus was related to a heterozygous prothrombin G20210A polymorphism associated with a

homozygous 4G/4G polymorphism of the plasminogen-activator-inhibitor. Jalal et al report a challenging case in which the rapidly evolving novel method of 3-dimensional printing was used to allow successful implantation of a Hybrid Melody valve through a transeptal approach via right atriotomy in a 4-month-old child with an atrioventricular canal abnormality and 3 previous failed surgeries.<sup>27</sup>

With all the medical issues surrounding the management of CHD, the associated psychosocial burden receives relatively limited attention. Patients with CHD have increased incidence of anxiety, depression, and other psychiatric/psychosocial problems.<sup>28</sup> These may lead to limited physical activity levels, which have deleterious effects on both mental and physical health. Cardiac rehabilitation can be an important tool in combating these issues but presents particular challenges in the CHD population.<sup>29</sup> In addition, it might be possible to combat these limitations with active psychosocial intervention. Kovacs et al provide the findings of a pilot study for a randomized controlled trial assessing the effectiveness of psychosocial intervention, showing both the feasibility and potential promise of this approach.<sup>30</sup>

Moving forward, we plan to continue the emphasis on CHD and pediatric cardiology in *CJC*. A theme issue on adult CHD is planned for the fall of 2019 and, in addition to state-of-the-art review articles in the area, will feature a CCS Guidelines report in the area.

## Conclusions

*CJC* is growing and succeeding. With the addition of *CJC Open*, the scope and range of CCS publications has expanded. We look forward to further expansion in congenital heart disease, pediatrics, and other rapidly developing and exciting areas in cardiology.

## Funding Sources

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## Disclosures

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