Canadian Journal of Cardiology 32 (2016) 8–12

Viewpoint

Multidisciplinary Aortopathy Clinics Should Now Be the Standard of Care in Canada

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ABSTRACT

Thoracic aortic aneurysm is often undiagnosed and has a very poor prognosis when presented with acute aortic dissection. Early diagnosis, expert medical management, and elective aortic surgery are the cornerstones of improvement of long-term survival in thoracic aortic disease (TAD). International guidelines now recommend the acute and long-term management of patients with TAD to occur within multidisciplinary aortopathy clinics under the care of professionals with specific training and experience. Multidisciplinary "heart teams" are recognized to be more focused on patient-centric care, to facilitate faster clinical decision times with increased adherence to guideline-directed therapy, and to improve knowledge translation and physician and patient satisfaction. The range of differential diagnoses for TAD has expanded rapidly over the past decade. Diagnosis of an index case with a syndromic or nonsyndromic familial TAD allows for preventative care. Effective family screening can save lives by allowing for

RÉSUMÉ

L'anévrisme aortique thoracique est souvent sous-diagnostiqué, et le pronostic en cas de dissection aortique aiguë est très sombre. Un diagnostic précoce, une prise en charge médicale experte et une chirurgie de réparation aortique préventive constituent les clés de l'amélioration de la survie à long terme des patients atteints d'une telle pathologie. Les lignes directrices internationales prônent désormais une prise en charge aiguë et à long terme par une équipe multidisciplinaire en clinique spécialisée. Les équipes multidisciplinaires « du cœur » ont en effet la réputation d'offrir des soins plus axés sur le patient, de prendre des décisions cliniques plus rapidement, de mieux suivre les lignes directrices de traitement et de faciliter la transmission de la connaissance, en plus d'améliorer le taux de satisfaction des patients et des médecins. La gamme des diagnostics différentiels en matière de pathologie aortique thoracique a pris beaucoup d'ampleur au cours de la dernière décennie, et un

Thoracic aortic aneurysm (TAA), although often undiagnosed, has an estimated incidence of approximately 10 per 100,000 person-years and acute aortic dissection (AoD) approximately 3 per 100,000 person-years.[1] Acute AoD currently has a very poor prognosis with 40% of patients who die immediately, 1% per hour who die thereafter, 5%-20% who die during or shortly after surgery, and only 50%-70% of survivors alive 5 years after surgery depending on age and underlying etiology.[1] Early diagnosis, expert medical management, and elective aortic surgery before AoD are the cornerstones of improvement of long-term survival in patients with thoracic aortic disease (TAD).

This theme issue of the *Canadian Journal of Cardiology*, on recent advances in understanding and treating TAD, provides reviews on new genetic discovery, better understanding of the structure and function of the aortic wall, and current recommendations for the medical and surgical management of TAD. Institutional multidisciplinary pathways developed for other cardiovascular emergencies such as ST-elevation myocardial infarction or acute stroke have not generally existed for acute AoD, but are now recommended in international guidelines.[1] Acute and long-term management of patients is also now recommended to occur in multidisciplinary aortopathy clinics under the care of professionals with specific training and experience with TAD.[2,3]

This 'Viewpoint' represents our personal experience of > 10 years in developing multidisciplinary aortopathy clinics in a pediatric and adult cardiology clinic setting, with the knowledge of the issues faced by similar clinics across Canada. The current goals of these clinics are presented with reference to the 2010 American guidelines,[1] the 2014 European guidelines,[2] and the 2014 Canadian Cardiovascular Society Position Statement[3] on the management of TAD.

Multidisciplinary Models of Care

Multidisciplinary care has a long history in the management of patients with chronic cardiovascular conditions. As

Received for publication September 21, 2015. Accepted October 7, 2015.

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See page 11 for disclosure information.

http://dx.doi.org/10.1016/j.cjca.2015.10.003

elective management of thoracic aortic aneurysm rather than emergent care of acute aortic complications. Expert cardiac imaging with access to the full range of required imaging modalities is central to all clinical management decisions. Medical and surgical management of TAD is now provided as personalized care according to patient- and disease-specific factors. Special considerations apply to pregnancy management for women with TAD. Multidisciplinary aortopathy clinics should now be the standard of care for the management of TAD in Canada and we should implement best practice guidelines. With the already established and emerging clinics, the stage is now set to build a Canadian Aortopathy Clinics Trials network.

patient qui reçoit un diagnostic de pathologie aortique thoracique syndromique ou non syndromique peut désormais obtenir des soins préventifs. Un dépistage efficace effectué auprès des familles exposées à un risque permet aussi de sauver des vies, notamment en permettant le traitement de l'anévrisme avant l'apparition de complications aiguës. Toutes les décisions cliniques doivent bien sûr reposer sur des résultats d'examens d'imagerie médicale de pointe, et la prise en charge médicale et chirurgicale des pathologies aortiques thoraciques doit s'effectuer en fonction du profil du patient et des particularités de sa maladie. Cela est notamment le cas pour le suivi de grossesse des patientes atteintes de cette affection. Au Canada, la prise en charge de la pathologie aortique thoracique devrait désormais toujours se faire dans une clinique multidisciplinaire spécialisée, et des lignes directrices de meilleure pratique clinique devraient être établies. Grâce aux cliniques déjà en place et à celles sur le point d'entrer en fonction, le temps est maintenant venu de créer un réseau de cliniques spécialisées dans la tenue d'études cliniques sur l'aortopathie.

the complexity of management increases with more evidence-based data and new interventional therapies, multidisciplinary "heart teams" have been identified to be more focused on patient-centric care. For example, these "heart teams" have now emerged as class I recommendations from the American and European professional societies for clinical decision-making with regard to coronary revascularization and transcatheter aortic valve replacement.[4] Historically, the "heart teams" in these settings have focused on the interventional cardiologist and cardiovascular surgeon, but have now been expanded to include imaging specialists, cardiac anaesthesiologists, intensivists, nurses, social workers, and designated clinical and/or research coordinators.[5] Expansion of this concept has seen the creation of multidisciplinary joint clinics, in which these teams now routinely see the patients at the same time. The real and perceived benefits of this multidisciplinary "heart team"-based approach include increased adherence to guideline-directed therapy, reduced clinical decision-making times, continuity of patient care, improved knowledge translation to patients and referring physicians, improved patient satisfaction and quality of life, improved physician satisfaction through opportunities for professional development, and opportunities for collection of data for research and more effective resource utilization.[5] Faster clinical decision times have been shown to occur in multidisciplinary clinics, as opposed to a multidisciplinary team meeting after a visit to a sole clinician.[6] Patients have also been shown to prefer multidisciplinary care. In a systematic review of randomized controlled trials with interventions including team-based and nonteam-based care, increased patient satisfaction occurred with team-based care.[7]

Expanding Differential Diagnosis

The range of differential diagnoses for TAD has expanded rapidly over the past decade. Since Marfan syndrome (MFS) was first described in 1896 and the *FBN1* gene discovered almost 100 years later in 1991, aortopathy genetic testing panels have grown to now include up to 20 genes in which mutations are known to cause TAD. Other related connective tissue disorders such as Loeys-Dietz syndrome (LDS), due to mutations in the *TGFBR1*, *TGFBR2*, *SMAD3*, *TGFB2* and *TGFB3* genes, also feature aortic root and ascending aortic involvement, but also more widespread arterial aneurysms and in some families significantly more aggressive disease with AoD that occurs even in early childhood. Vascular Ehlers-Danlos syndrome (vEDS) due to *COL3A1* mutations might have aortic involvement, but features more medium-sized arterial involvement including coronary, splanchnic, and uterine artery dissection and rupture. Familial TAA and dissection (FTAAD) can be due to mutations in some of the genes mentioned without other major systemic features of a connective tissue disorder, but also due to other genes including *ACTA2*, *MYH11*, *MYLK*, and *PRKG1* involving vascular smooth muscle cell function in the aortic wall. Genetics also plays a role in congenital heart diseases such as bicuspid aortic valve, tetralogy of Fallot, and transposition of the great arteries, and in association with other congenital syndromes like Turner syndrome, which can all be associated with TAD.

Importance of Screening

TAA patients are most often asymptomatic and the diagnosis is usually made after imaging for other reasons or for screening purposes. Having a genetic counsellor in the multidisciplinary aortopathy clinic setting to assist in performance of pedigree analysis and family screening is invaluable. Clarification in the family history with regard to actual location of any aortic or arterial aneurysm in other family members, the specific nature of any "sudden deaths," and the availability of any confirmatory autopsy report is essential. Diagnosis in the index case is made on the basis of clinical findings and exclusion of known genetic syndromes, targeted imaging and genetic testing tailored to the patient, and family history of vascular events.[1-3] Ideally, a full clinical examination of the patient is performed by a medical geneticist with expertise in connective tissue disorders before a decision on which genetic test(s) to order. When a familial TAD is suspected, genetic counselling can be offered and genetic testing requested accordingly. Because of the autosomal dominant transmission of these