BC Inherited Cardiac Conditions and Sudden Cardiac Death Management

Prepared For: PHSA and MOH Senior Leaders
For: Discussion
By: Heather Jackson
Date: January 15, 2019

WHAT IS THE PROBLEM WE ARE TRYING TO SOLVE?

Up to **2.56% (123,319 individuals) of British Columbians** are at risk of having an Inherited Cardiac Condition (ICC)[1] and are predisposed to Sudden Cardiac Death (SCD). If risk is recognized and managed early, the consequences of **SCD can be prevented**; however, these conditions do not always present with symptoms, and for many patients/families, the first sign of the condition is when someone dies suddenly. Collaboration and coordination among the practitioners responsible for the assessment of the individual who died of SCD/suspected ICC, and the clinical management team of the surviving family, is essential to prevent further death.

International guidelines published in 2011[2] and 2013[3] state that best practice for ICC/SCD individuals and families is management by interdisciplinary care teams in specialized clinics, integrating expertise of cardiology, genetics, genetic counselling, and pathology (Class 1 recommendation). Furthermore, a cardiac autopsy with tissue and/or blood retention at the time of autopsy for all SCD/suspected ICC cases, and referral for the clinical evaluation of all first-degree relatives are also Class I recommendations[4]. **These guidelines are not consistently practiced in BC,** limited by system complexity, operational logistics, and insufficient and/or uncoordinated human resources.

In the current state, it is *possible* for patients and families to find fragmented support throughout the province. Their success, however, depends on: 1) their diagnosis, 2) their geographical location, 3) their existing access to care, and 4) their health care providers' knowledge and awareness of these services and the underlying condition.

In BC, isolated, niche clinics within cardiology and genetic disciplines have organically evolved. Independently, they have attempted to expand their service over the past five years to meet the growing demands of these patient populations. However, without a common strategy, provincial human

---

[1] ICCs include: Inherited arrhythmias, cardiomyopathies, heritable aortopathies, familial lipid disorders, pulmonary hypertension

[2] Ackerman, M.J. et al (2011) HRS/EHRA Expert Consensus Statement on the State of Genetic Testing for the Channelopathies and Cardiomyopathies. *Heart Rhythm* 8, 1308-1339

[3] Prior, S.G. et al (2013) HRS/EHRA/APHRS Expert Consensus Statement on the Diagnosis and Management of Patients with Inherited Primary Arrhythmia Syndromes. *Heart Rhythm* 10 (12)

[4] Semsarian, C. Ingles, J. and Wilde, A. A.M. (2015) Sudden cardiac death in the young: the molecular autopsy and a practical approach to surviving relatives. *European Heart Journal* 35, 1290-1296.

resource plan, or centralized infrastructure, fragmented care has emerged and has led to inefficient, inconsistent and inequitable service.

Increasing public awareness and understanding of genetics, patients' expectations regarding appropriate availability of genetic testing, growing provider awareness and expertise, expanding technological innovations and research, and overall knowledge dissemination of cardiac genetics is driving **unprecedented growth and demand in this area of health care.** A comprehensive, holistic provincial program is imminently needed to simplify care pathways for individuals and families of ICC/SCD, and to minimize frustration and lack of clarity in practioners attempting to provide and/or access this service. This will not only lead to the potential avoidance of sudden cardiac death for BC patients, but it will also result in reduced clinical redundancy, improved efficiency in resource allocation, and enhanced and standardized support for families. Collaborative program development work will help establish better cross-sectoral stakeholder partnerships, which can serve as a necessary foundation for this rapidly evolving area of medicine.

WHY NOW?

1) Under its new mandate from the Ministry of Health (to oversee provincial clinical coordination, clinical policy development and clinical standardization), and with oversight of Medical Genetics, Provincial Lab Services, BC Children's Hospital (BCCH), and Cardiac Services BC (CSBC), PHSA is well positioned to lead the coordination and development of a BC Inherited Cardiac Conditions and Sudden Cardiac Death Management Service.
2) Guidelines defining appropriate management of ICC and SCD were defined in 2011 and 2013 and BC has yet to implement the necessary programmatic changes to deliver this service in a coordinated and efficient manner.
3) The establishment of a personalized medicine portfolio at the Ministry of Health would indicate that this service is in line with the future direction of health care policy in the province.
4) Currently, there is collective momentum and consensus among the key stakeholders of ICC/SCD to move towards a more holistic approach, with a common goal of closing the care gaps and preventing any further unnecessary SCD in BC.

PROPOSED SOLUTION:

The ideal program will function as a province-wide, centrally-coordinated network for referral and case management of all inherited cardiac condition (ICC) patients and families associated with sudden cardiac death (SCD). It should leverage existing infrastructure, provide comprehensive clinical evaluation and diagnostics, and enable timely access and communication for patients/family members, regardless of condition, age and geographical location. Because of the direct link between ICC and SCD, strong partnerships among a broader system of care (provincial laboratory medicine, pathology, the BC Coroner's Office, cardiology and medical genetics) is required. Streamlining services will not only support a family-centered model of care across geographical locations, but will also enable patients to effectively flow from one clinical service to another. Furthermore, a unified program will strengthen

accountability, transparency, monitoring, reporting, and knowledge sharing through its cross-sectoral multi-stakeholder partnerships, enhancing care for patients and families laden with these diagnoses. A comprehensive and holistic approach is essential to avoid unexpected death and to minimize the impact that ICCs and SCD have on patients and families living in British Columbia.

WHAT IS THE CURRENT RISK OF NOT CHANGING?

Perspective	Risk/Consequence
Patients & Families	▪Unnecessary death due to lack of risk identification, referral and/or clinical care ▪Death on an existing wait list due to long wait times caused by insufficient resources[5] ▪Not receiving evidence-based, standard of care ▪Inability to pursue genetic testing of a deceased relative at a deferred time (when families are emotionally ready) due to system-wide, inconsistent policies/protocols in tissue retention at the time of death ▪Inequitable access to services based on geographical location, due to many contributing factors ▪Inconsistent messaging of the significance/interpretation of genetic findings ▪Confusion – who to contact ▪Mismanaged transitions of care; between primary care and specialized service and among specialists involved in SCD/ICC ▪Disconnect between adult and pediatric services within the same family; disconnect between coroners and the clinical system when dealing with a death of a family member ▪Communication breakdown; delayed communication ▪Insufficient communication materials for the dissemination of genetic results among family members and explanation of familial risk ▪Lack of sufficient psychological and social support to manage decisions, interpret findings and incorporate necessary lifestyle modifications into daily living ▪Lack of sufficient pediatric and family support to ensure sufficient resources are available to manage lifestyle/activity restrictions in children ▪Financial burden with respect to costs, time, travel ▪Work and social absenteeism for in-person visits when virtual service is a proven alternative for components of care
Health Care Providers	▪Inability to align work with professional guidelines (evidence-based practice) ▪Lack of coordination among disciplines to ensure comprehensive and consistent management of ICC/SCD families ▪Missing communication/documentation ▪Misinterpretation of clinical significance of genetic findings due to increasing complexity of results and lack of training/expertise in a rapidly evolving field (results are probabilistic, not deterministic) ▪Moral distress due to not being able to see patients in a timely manner when risk

[5] This has already occurred in the current system; SCD of a patient referred to an existing program

	of death can be imminent[5] ▪Redundant documentation due to silos of care and IT barriers ▪Providers not working to full scope of practice OR forced to work out of scope of practice to fill a clinical need ▪Workload burden and staff burnout
Health System	▪Silos of care; redundant and inefficient services ▪Managing multiple competing proposals for similar/overlapping services due to lack of coordination ▪Inappropriate health care utilization and resources ▪Inefficient use of genetic testing ▪Delayed decisions for approving genetic testing ▪Lack of accountability and policy direction ▪Significant current unmet need; lack of future sustainability ▪Lack of human resource planning to meet demands of current or future state ▪Escalating potential of risk with system-wide inaction to plan/deliver guideline-based care

WHAT IS THE FUTURE RISK IF WE DON'T CHANGE TODAY?

- Unnecessary sudden cardiac death in BC, potentially occurring in the same family more than once
- Missed opportunities to effectively identify the genetic basis of SCD and deaths suspected to be ICC today may lead to missed opportunities of saving a life in the future
- Cardiac genetic testing results will only continue to increase in complexity (next generation sequencing; whole genome sequencing) and will require greater collaboration among disciplines to interpret clinical significance of findings and determine the optimal/appropriate clinical care plan for patients and families; a strong provincial foundation is required to effectively manage the system of care on the horizon.
- Inability for the clinical system to adapt to advancing technologies in genetics/genomics due to the lack of centralized management
- With increasing availability of genetic testing and lowered costs, there is a potential for testing to become a standard diagnostic tool ordered by general cardiology/internal medicine/primary care physicians. There is a significant risk of inappropriate ordering of tests (wrong patient or wrong test) or the misinterpretation of genetic findings and resultant mismanagement of patients and families. Genetic testing is deemed to be probabilistic, not deterministic, and the interpretation of complex findings will require clinical interpretation to establish if and how results should be incorporated into cardiac management plans. Once this service is distributed, it will be harder to ensure consistent and appropriate use of testing.
- Lack of sufficient skilled professionals to meet the service demand of the province due to lack of human resource planning and financial impact assessments

GUIDING PRINCIPLES FOR PROGRAM DEVELOPMENT

1. Evidence-Informed: Safe, Appropriate, High Quality
2. Family & Patient-Centered

3. Accessible, Timely & Comprehensive
4. Integrated & Coordinated Across System (including primary care)
5. Sustainable, Efficient & Cost Effective
6. Innovative, Visionary & Scalable (to meet changing genetic landscape)
7. Collaborative & Supported by Strategic Partnerships
8. Ethical, Transparent
9. Strategic alignment with MOH, IHI and PHSA

STRATEGIC ALIGNMENT:

1. MOH: Quality, safe, timely, cost-effective, collaborative, team-based care, coordination between Primary and Specialized care, innovative and sustainable solutions, optimizing scope of practice, effective resource utilization
2. IHI: Patient-Experience, Provider-Experience, Sustainable, Population Health
3. PHSA: Province-wide solutions for excellent care, dare to innovate, cultivate partnerships.
4. BC Quality Matrix: Acceptable, Appropriate, Accessible, Safe, Effective, Equitable, Efficient

NECESSARY NEXT STEPS:

1) Due to the current distribution of services affiliated with this proposal, a collective discussion with PHSA Senior Leaders is necessary to identify and approve operational changes required to deliver an integrated service. Required leaders include those with oversight of:
    - Cardiac Services
    - Medical Genetics
    - Lab Services and Pathology
    - Children's Hospital
2) Engagement with MOH Specialized Programs, Out of Province Testing, and Personalized/Precision Medicine portfolios
3) Discussions with the BC Coroners Service to identify improvement opportunities to align SCD management with existing guidelines
4) Engagement with HAs to understand how to serve as a true provincial program distributed among the HAs
5) Engagement with primary care to understand the ideal transitions for these patients and families
6) Strategic stakeholder meeting including senior decision makers and key opinion leaders – Moving Plan to Action
7) Consensus/Finalization of Program Scope, Operational Impact Assessment and Implementation Proposal