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Editorial

A National Call to Action—Improving the Detection of Critical Congenital Heart Disease

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See article by Wong et al., pages 199–208 of this issue.

Congenital heart disease is common, detectable, and treatable, with excellent outcomes in the modern era. However, in this nation we still "miss" too many cases of critical congenital heart disease that are detectable in newborns before they decompensate. In most cases, decompensation relates to closure of the ductus arteriosus. In the fetus, in whom gas exchange occurs in the placenta, the ductus arteriosus serves to divert most of the right ventricular cardiac output away from the high-resistance pulmonary vascular bed to the low-resistance placental circulation. After birth, newborns with critical left-sided cardiac obstruction are dependent on blood flow from the right ventricle through the ductus arteriosus to support the systemic circulation, whereas those with critical right-sided cardiac obstruction are dependent on the ductus arteriosus to maintain pulmonary blood flow. Therefore, between birth and ductal closure, neonates are often clinically well; hence, this period is our opportunity to make the diagnosis, optimize their medical condition, and prepare them for timely definitive intervention.

Screening using oximeters has been recognized as a potential approach to improve our detection rates given that many forms of critical heart disease are hypoxemic in the neonatal period (Table 1). Although many patients with critical congenital heart disease are diagnosed antenatally or in the early postnatal period during routine clinical evaluation, there remains a "diagnostic gap," in which current practice does not detect patients with critical congenital heart disease before cardiovascular decompensation. In this issue of the *Canadian Journal of Cardiology*, Wong et al.[1] present evidence that newborn oximetry screening can reduce the diagnostic gap for newborns with critical congenital heart disease. In a collaborative process involving members of the Canadian Pediatric Society, the College of Family Physicians, and the Canadian Cardiovascular Society, they have made important recommendations that recognize the value of a comprehensive iterative assessment including prenatal ultrasonography, physical examination, and oximetry screening in newborns. The incremental value of newborn oximetry screening is tremendously important; however, we must note that this form of screening does not capture all cases of critical congenital heart disease (a recent systematic review reported a sensitivity of 76.5%[2]; the most common false-negative result has been in newborns with coarctation of the aorta). Hence, newborn pulse oximetry screening should be used as an adjunct to current clinical practice; it should not replace prenatal ultrasonography or the newborn physical examination.

The implementation of policies that reflect this opportunity to improve child health is the next step. At the provincial level this initiative needs to be supported, while we must recognize that at the regional level the implementation may take on various forms. We are a geographically large nation with centralized expertise in pediatric cardiology at relatively few institutions. As such, the referral centres need to work in concert with local partners to develop a practical approach during the implementation phase. The Canadian Pediatric Cardiology Association has an active and engaged membership that is eager to work collaboratively with health care partners in this process. We are behind other developed nations including the United States, Australia, and the United Kingdom in policy development and implementation and consequently, every day we continue to put newborns at risk.

In the past year, I have personally cared for 4 children who would have benefitted from newborn screening. As a case in point, I was referred a young child who received all her prenatal, postnatal, and childhood care here in British Columbia. She went for a dental procedure at a local hospital at 4 years of age and was noted to be desaturated. Upon referral, she was diagnosed with transposition of the great arteries. In 2016 with modern medical care, transposition of the great arteries should be universally diagnosed antenatally or in the immediate newborn period. Indeed, my patient was lucky to survive because the natural history of this condition is such that newborns have an average life expectancy of 0.65 years without intervention,[3] and many times children with missed

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Table 1. Examples of critical congenital heart disease

Critical coarctation of the aorta
Double-outlet right ventricle
d-Transposition of the great arteries
Hypoplastic left heart syndrome
Interrupted aortic arch
Pulmonary atresia
Total anomalous pulmonary venous return
Tetralogy of Fallot
Tricuspid atresia
Truncus arteriosus

critical congenital heart disease are diagnosed only at autopsy. Although the survivors I have seen are the lucky ones, their surgical courses were all modified because of late diagnoses compromising their chances for an excellent long-term outcome—something we would currently expect for newborns with these conditions. Interestingly, the common thread shared by the patients I have seen with missed critical congenital heart disease is social vulnerability. These were children of immigrants living in homes in which little or no English was spoken. We know social vulnerability clusters with other risk factors that conspire to limit children routinely accessing health care. Consequently, screening programs that occur during one of the few obligatory interactions with the health care system—childbirth—afford us an opportunity to identify these newborns when they are medically stable.

We must also recognize the challenges associated with newborn oximetry screening. In a country as geographically vast as Canada, specialized services like pediatric cardiac consultation and pediatric echocardiography are not locally available for many health care practitioners. Transportation of neonates is time-consuming and costly and carries inherent risk for specialized transport teams. The judicious use of these resources is always a consideration. Pediatric cardiologists across the country will need to work with local communities to jointly plan the most appropriate triage process for those newborns who are identified based on newborn oximetry screening. Given the potential for rapid clinical deterioration, neonates who are identified based on newborn pulse oximetry screening require prompt definitive diagnosis and treatment. This potential for early and rapid clinical decline sets this apart from many other forms of newborn screening. Nonetheless, there are several examples of successful implementation of newborn oximetry screening from other jurisdictions that may serve as a model for practitioners across Canada.[4] As we now move toward implementation of newborn pulse oximetry screening, we need to work with our partners in health care, hospital administration, and provincial ministries of health to ensure timely local implementation that is effective and also to evaluate the impact of this initiative on health outcomes and resource utilization.

Disclosures

K.C.H. is the president of the Canadian Pediatric Cardiology Association.

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