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EDITORIAL

Congenital Heart Disease Requires a Lifetime Continuum of Care: A Call for a Regional Registry☆

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Introduction

Congenital heart disease (CHD) is the most common congenital disorder in newborns [1]. Current global estimates suggest nine babies are affected in every 1,000 live births corresponding to 1.35 million babies born with some form of CHD each year [1]. Within Australia it is estimated that CHD affects over 2,400 babies each year; half of whom will require surgery or catheter-based intervention [2]. Current Australian 'true' prevalence rates, based on the Netherlands National CHD registry, indicate there could be around 32,000 patients under the age of 18 years with CHD in Australia. A similarly calculated corresponding figure for New Zealand is estimated to be approximately 10,000. Taking into consideration newborns through to adults living with this disease, the total CHD population could conceivably represent well over 65,000 Australians. Many patients have minor lesions, but there is an imperative to understand the long-term outcome for those who have required CHD intervention or are identified as being at significant risk for complications.

Contemporary surgical procedures have improved the outlook for CHD patients such that the majority (>95%) of patients are now expected to survive into adulthood [3,4]. Despite this success, CHD continues to be a major global health burden. Key public health challenges include lack of knowledge on risk factors for CHD, its prevalence across the lifespan, and long-term outcomes for adults with CHD, and their impacts in terms of health-care provision [5]. Improved understanding of the total numbers of patients living with CHD and the total burden across the disease continuum is imperative for patients and for those responsible for treating them, including health policy makers. Here we discuss how an Australian and New Zealand Regional CHD Registry can bring us a step closer to achieving this goal.

Unknowns in CHD Epidemiology

Incidence and prevalence data of childhood CHD derived from small subpopulation registries vary across the globe [1,6]. The Dutch CONgenital CORvitia (CONCOR) national registry, initiated in 2002, now includes almost 15,000 patients from 102 hospitals and is, to date, the largest CHD registry in the world. However, this registry may capture only one-third of all expected CHD patients and one-fifth of patients with severe CHD, as revealed by application of the empirical prevalence data derived from the population-wide cohort in Quebec, Canada (birth prevalence rates of 11.89 cases per 1,000 children and 4.09 cases per 1,000 adults within the population of patients hospitalised for CHD) [6,7]. Thus, our current understanding of the epidemiology of CHD is limited by the data available to inform it. We have a unique opportunity within the Australian/New Zealand landscape to significantly address this knowledge gap.

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Changing Survival Trends: A Story of Surgical Success

Historically, survival of CHD patients with significant lesions beyond early childhood was limited, [4] with less than one-third of infants being expected to survive into adulthood [3]. Major advances have been made both in the available surgical techniques (there are more than 150 surgical procedures in CHD [8]) and in the recognition of the need for early surgical intervention.

The first 12 months of life are the most critical for survival; 96% of infants surviving beyond this milestone are expected to survive to adolescence [6]. Analysis of data from Canada has established, over time, a marked change in the distribution of age at death with a shift towards deaths at older ages and temporal changes in mortality with a 15-year increase in the median age at death [4]. For the first time in history, the static birth prevalence and the significant reduction of operative mortality for surgical CHD interventions have given rise to a greater number of adults than children living with CHD.

This changing demographic and epidemiology of CHD has shifted a large portion of the burden of disease into the adult population, with little understanding of the cost to patients, the health system, and the community. After linkage of the CONCOR registry with the Dutch national mortality registry, data interrogation revealed that the majority of deaths in CONCOR patients (n=197) were of cardiovascular origin (77%) and that almost half were due to arrhythmias or progressive heart failure [9,10]. It also demonstrated that adult patients with CHD have excess mortality compared to the general population.

Implications for the Continuum of Care

Ongoing research is required to help better understand normal cardiac development and to answer questions about how it can go wrong. Much progress has been made in our understanding of the genetic and environmental contributors to CHD, which in turn have helped inform inheritance patterns and the risks of recurrence [2]. Recent research demonstrates the benefits of integrating psychological support and genetic counselling into routine CHD care, including improved psychosocial well-being amongst parents [11].

The management of adult CHD is associated with an increasing prevalence of complications arising from haemodynamic or hypoxic stress, postoperative sequelae, residual defects and acquired comorbidities [3,12]. It has been long known that repaired conditions such as tetralogy of Fallot mandate ongoing follow-up because of a potential need for reoperation. We are only now beginning to understand that childhood surgical repair of even the most benign CHD conditions, such as ventricular septal defect (VSD) and atrial septal defect (ASD), can negatively impact on ventricular function and exercise capacity in adulthood [13–15].

Significant progress has been made in the development of surgical techniques and the tissues used in CHD repair strategies. However, we need to be cognisant of the potential impacts and longer-term outcomes of the tissues being used. Prosthetic materials may be life-saving, but they can be limited by durability, infection risk, host-immune responses and thrombotic complications. Longer-term follow-up is needed to fully understand the health-related implications of these limitations. Tissue engineering is emerging as a promising strategy to better enable individualised therapy that affords the benefits of regeneration, remodelling and growth potential [16].

Additionally, there is a lack of knowledge as to the overlaying impact of congenital heart defects, repair strategies and chronic conditions (e.g. hypertension, obesity) on the health of other body systems and organs. It is vital that ''beyond survival'' outcomes are taken into account. A key aspect of such care includes improved understanding of long-term physical, reproductive and psychosocial impacts of CHD on adults [5].

Gaps in Care

Childhood CHD is managed via specialist services with ongoing patient follow-up. Ideally, at the age of 16-18 years these children should be transitioned to an adult CHD service. Even after successful treatment, many patients require life-long cardiac surveillance [17]; however, loss to appropriate cardiology follow-up can be substantial and this lapse in care may put patients at increased risk of adverse outcomes [18,19].

Canadian data have demonstrated that 61% of CHD patients, including 79% of those with severe CHD, fail to receive cardiac follow-up after their 18th birthday despite still receiving regular primary physician care [20]. More recently, US-derived data show that 42% of adults with CHD had gaps of greater than three years in their cardiology care; typically commencing at age 19 years (during the transitional period to adult care), the gap lasted longer than 10 years for 8% of patients [18]. Return to care was prompted by symptoms, healthcare professional referral, or a desire to prevent potential problems. In the UK this figure is lower (24%) and occurs later (median age 32 years), but the duration of the loss to follow-up is longer (median duration 22 years) [19]. Notably, the UK data show that almost half of all subsequent deaths in CHD patients occurred whilst they were not under specialist care. Contemporary Australian data have demonstrated alarming rates of delay in treatment for pulmonary hypertension within the CHD population [21].

Adequate planning of transition care, particularly for postoperative patients and those with a recognised risk of late complications, and the establishment and maintenance of comprehensive regional adult CHD services are clearly needed. However, evidence supporting the benefits of effective transitional care on long-term patient outcomes in adults