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Original Article

Landmark lecture in nursing: a life-cycle perspective on CHD: What happens beyond your clinic?*

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Abstract Over the past decades, survival of patients with CHD improved significantly, making it a life-cycle disease. Hence, there is a need for a workforce that can take up the care for afflicted individuals in the different phases of the life spectrum. Each life phase is associated with specific challenges. Topics that should receive more attention in clinical care or in CHD research are parenting styles of parents of children, transfer and transition of adolescents, cumulative burden of injury in the brain in adults, and geriatric care for older persons with CHD. Nurses, along with other healthcare professionals, will play a pivotal role in building up expertise in these areas and taking up these challenges.

Keywords: Heart defect; congenital; interdisciplinary; transition; parenting

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It is well known that the spectacular improvements in medical and surgical therapies for children born with CHD yielded an increased life expectancy. To date, >90% of the children with CHDs can survive into adulthood.[1,2] Due to these favourable evolutions, CHD can be seen as a life-cycle disease. Hence, patients and families are in need for a workforce that can provide expert care in each stage of life, from fetus to older person. Newborns and children receive care and follow-up at paediatric cardiology departments. For the growing group of adults with CHD, dedicated adult CHD programmes have been established.[3] Nurses and Advanced Practice Nurses play a pivotal role in the interdisciplinary care for individuals with CHD, throughout the life spectrum.[4–8]

At the 7th World Congress of Pediatric Cardiology and Cardiac Surgery, held in Barcelona (Spain) from 16 to 21 July, 2017, a series of Landmark Lectures was given. The Landmark Lecture in Nursing was entitled "A life-cycle perspective on congenital heart disease: What happens beyond your clinic?". The present article describes the four topics that have been addressed in this Landmark Lecture in Nursing, each of which was linked to a particular stage in life: childhood, adolescence, adulthood, and older ages. These presented topics require more attention both in clinical care and in research.

Childhood: parenting style and its consequences

Over the past decades, numerous studies have investigated what it means to parents to receive the message that their fetus has CHD,[9,10] or how their life is affected by growing a child with this condition.[11,12] Research on parenting style and its consequences, however, is sparse.

An assumption that is sometimes expressed by clinicians is that parents of children with CHD are overprotective, and therefore limiting the full potential of their child. This assumption has been tested for the first time in a cohort of 429 adolescents with CHD.[13] Using dedicated self-report scales, three

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parenting dimensions were assessed: regulation, psychological control, and responsiveness.[13] Regulation refers to the level of behavioural control. A sample item of this dimension reads, "My mother/father asks me questions about how I am behaving outside the home". Psychological control pertains to the level in which parents try to influence the emotions and thoughts of their child. A sample item reads, "My mother/father is always trying to change how I feel or think about things". Responsiveness is the level of support that children experience from their parents. An example of an item is, "My mother/father makes me feel better after talking over my worries with her/him". The combination of these three parenting dimensions determines the parenting style. Four meaningful parenting styles could be identified in parents of patients with CHD: democratic (27%), overprotective (30%), indulgent (26%), and coercive (17%).[13] This distribution of parenting styles was not significantly different from the distribution among parents of healthy youngsters.[13] These findings refute the notion that parents of children with CHD are more overprotective than parents of healthy children.

The second aim of that study was to investigate whether parenting styles were associated with patient-reported outcomes.[13] It was found that parenting styles, indeed, were associated with depressive symptoms, loneliness, quality of life, health status, alcohol use, binge drinking, smoking, and illicit drug use. More specifically, patients who experienced the parenting style as authoritative (= democratic) had the most favourable outcomes, and patients who experienced their parents as being overprotective or coercive had the worst outcomes.[13] These two latter parenting styles are characterised by a high level of psychological control, which clearly is detrimental. On the other hand, a high level of behavioural control and support is beneficial. These effects have been confirmed in a subsequent longitudinal study.[14]

Another study, conducted on 192 adults with CHD, has investigated whether parental overprotection recall was associated with current heart-focussed anxiety.[15] A moderate positive correlation ($r = 0.30$; $p < 0.001$) between the two variables has been found.[15] Thus, the study also confirmed that overprotection is detrimental for future functioning.

Regarding parenting style during childhood, it can be concluded that

- Parents of patients with CHD are not more overprotective than parents of healthy children.
- Parenting style impacts on health and psychosocial outcomes:
  - o Parental support is good.
  - o Behavioural control is good.
  - o Psychological control is bad.
- Coaching and guidance of parents should include empowerment to develop a democratic parenting style.

Adolescence: transfer and transition

Throughout childhood, patients with CHD are treated and followed-up at paediatric cardiology. When they become adults, it is advocated that their medical surveillance and follow-up care be transferred to adult-focussed facilities.[16–19] Transfer is defined as "An event or series of events through which adolescents and young adults with chronic physical and medical conditions move their care from pediatric to an adult care environment".[20] A seamless transfer between paediatric and adult settings warrant that age- and developmentally appropriate care is provided, while assuring that patients remain under follow-up.[21] Research, however, has indicated that a substantial number of patients do have gaps in their care. In patients with CHD, care gaps rates range from 7 to 76%,[22] with a median proportion of 42%. The consequences of such care gaps may be far-reaching. Studies have reported that patients who presented for medical check-up after a care gap more often had a new diagnosis of haemodynamic significance and had a greater likelihood of needing an urgent surgical of catheter-based intervention.[23–25] Therefore, the identification of patients at risk for care gaps and the implementation of interventions that prevent such care gaps are utmost important.[26] Studies on risk factors of care gaps focussed only on patient-related factors. However, it is possible that hospital and healthcare system characteristics also play a role. Future studies, therefore, should investigate risk factors for care gaps from a multilevel perspective, scrutinising patient-related, hospital-related, and healthcare system-related factors. The Adole7C project aims to do so (www.adole7C.weebly.com).

Transition can be seen both as a developmental process and as a healthcare intervention. As a developmental process, transitions are passages from one life phase, physical condition, or social role to another, resulting in a temporary disconnectedness of the normal way of living, which demands an adjustment of the patient and the environment.[27,28] As a healthcare intervention, transition is defined as "a multifaceted, active process that attends to the medical, psychosocial, and educational/vocational needs of adolescents as they move from the child-focused to the adult-focused healthcare system".[29] In such an intervention, the adolescents are prepared to take charge of their lives and their health in adulthood.[20]

A Cochrane review published in 2016 analysed the effectiveness of structured transition programmes for young persons with chronic conditions.[30] The review included four controlled trials that were conducted on patients with spina bifida, CHD, diabetes, and different chronic conditions. The follow-up of the patients ranged from 4 to 12 months, and a total of 238 patients were included. The overall conclusion of this review was as follows: "… since few studies were eligible for this review, and the overall certainty of the body of this evidence is low, no firm conclusions can be drawn about the effectiveness of the evaluated interventions. Further research is very likely to have an important impact on our confidence in the intervention effect and likely could change our conclusions".[30] Therefore, more effectiveness studies on transition programmes are required. Currently, two trials are in progress. The first trial is the Chapter 2 study, which is a cluster randomised trial in two centres in Canada.[31] The transition intervention comprises two sessions of 1–1.5 hours each, focussing on patient education and self-management. Indeed, it is known that adolescents with CHD have gaps in their level of knowledge[32] and self-management.[33] The second trial is the Stepstones project, which is using a hybrid experimental design in seven centres in Sweden.[34] The transition programme in this trial includes eight key components: a transition coordinator; a written person-centred transition plan; provision of information and education; availability by telephone and email; information about and contact with the adult CHD clinic; guidance of parents; meeting with peers; and the actual transfer to adult care.[34] The primary outcome in this trial is the level of patient empowerment, as measured with the Gothenburg Young Persons Empowerment Scale (Acuna Mora et al, under review).

Regarding transfer and transition of adolescents with CHD, the following can be concluded:

- Care gaps are most prevalent in patients with CHD.
- Interventions to avoid care gaps should be implemented.
- Intervention studies on the effectiveness of transition programmes in CHD are in progress.

Adulthood: cumulative burden of injury in the brain

Neurodevelopmental problems in children with CHD are well known.[35] Research has identified multiple exposures that affect their neurocognitive functioning. In fetuses and newborns, genetic syndromes occurring in about 30% of people with CHD, epigenetic factors such as low birth weight or gestational age, and hypoxia due to altered cerebral perfusion are most prominent exposures.[36] The consequences are expressed as abnormalities in the brain structure, reduced brain volumes and microcephaly, and brain dysmaturation. Indeed, brain maturation slows in the 3rd trimester of the pregnancy.[37] Overall, 8–33% of newborns with complex heart defects that requires cardiac surgery are found to have impaired prenatal brain development.[35]

In children and adolescents, the complexity of the heart defect, cardiopulmonary bypass and perioperative hypoxia, abnormal haemodynamics, and cyanosis may injure the brain.[36] This results in altered white matter microstructure, neurodevelopmental anomalies, changes in behavioural outcomes, hampered educational achievements, and an increased risk for developing acquired brain injuries such as stroke[38] or periventricular leukomalacia.[39] The academic consequences of these neurodevelopmental challenges are demonstrated in a population-based study, which showed that children with CHD have a 50% higher likelihood to receive special education than those born without birth defects.[40]

An issue that has received little attention so far is the brain of adults with CHD. However, it is argued that there is a cumulative burden that yields additional injuries of the brain. Indeed, the accelerated development of atherosclerotic diseases, the higher incidence of diabetes, hypertension, atrial fibrillation, and heart failure are all morbidities of CHDs that affect the brain.[36] Hence, on top of the pre-existing neurodevelopmental burden since fetal life, the additional cerebrovascular lesion burden makes the brain more prone for developing acquired brain injuries and even early-onset dementia.[36] Neurocognitive decline is therefore becoming an emerging topic in CHD.

The cumulative burden of brain injury implies that healthcare teams in adult CHD should develop or involve expertise regarding neurocognitive functioning. Getting an understanding of the relative importance of the risk factors for neurocognitive disability and implementing interventions to promote brain health will be of paramount importance.[36] Nurses will be able to play a critical role in the improvement of patient-centred and societal outcomes by educating patients and their families, providing neurobehavioural interventions, and reducing the adverse effects of the heart defect on mental health.[36]

In conclusion, with respect to neurocognitive functioning, it can be stipulated that

- The heart–brain axis is getting increased attention.
- Neurodevelopmental issues in CHD are known.
- Additional cerebrovascular burden will play an important role in neurocognitive outcomes/decline of adults with CHD.

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Older ages: geriatric CHD

The term 'geriatric CHD' was used for the first time in 2011 to raise attention for the growing number of older persons with CHD.[41] Mortality data in patients with CHD have showed that the majority of the patients die when they are older than 60 years of age.[42] Indeed, of all patients who survive into adulthood, 90% of patients with mild, 75% with moderate, and 40% with complex heart defects survive to the age of 60 years.[43] With the contemporary life expectancy, the number of older persons with CHD is anticipated to grow markedly. By 2030, it is estimated that 11% of the adult CHD population is 60 years of age or older.[44]

The first empirical data show that older persons with CHD constitute a specific group of individuals.[41,45] Morbidity, healthcare utilisation, and mortality in this group of patients is high.[41,45] Therefore, the American Heart Association published a scientific statement on CHD in the older adult.[46] Nonetheless, little is known about the unique needs of these patients to date. The accelerated ageing process in persons with CHD, the comorbidities, and the changed responses to medication will yield significant challenges for healthcare professionals working with these patients. Moreover, the typical geriatric syndromes such as cognitive decline, immobility and falls, failure to thrive, and sensory alterations will emerge, and will necessitate competencies in geriatric care within the CHD teams. In addition, research should be undertaken on the altered functional and psychosocial profile of older persons with CHD to get an understanding of their health needs profile.

Concerning older persons with CHD, it can be concluded that

- Geriatric patients with CHD are a new and emerging population.
- Knowledge of specific needs of older patients with CHD is limited.
- Nurses and other clinicians should develop specific expertise in order to be prepared to take up the care for this growing and high-demanding group of patients.

Conclusions

Nurses and other healthcare professionals have to take up emerging challenges in the care for patients with CHD, and need to build up expertise in less established areas. Topics that should receive more attention in clinical care and in CHD research are parenting styles of parents of children, transfer and transition of adolescents, cumulative burden of injury in the brain of adults, and geriatric care for older persons with CHD.

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Conflicts of Interest

None.

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