Compendium C - National Strategic Action Plan for Childhood Heart Disease (CHD) 2019 Supporting Evidence

Table of Contents

Introduction and Identification of Priorities *2*

Linkages of priorities and focus areas with key policy documents *3*

CHD Prevalence and Impact *4*

Priority 1 Management, care and support *8*

Standards of Care 8

Infrastructure and Workforce 10

Priority 2 Supportive communities *12*

Awareness and Education 12

Neurodevelopmental and Mental Healthcare 13

Priority Populations 14

Priority 3 Research *16*

Research Priorities 16

Research Surveillance 18

References *19*

Introduction and Identification of Priorities

The National Strategic Action Plan for Childhood Heart Disease (CHD) 2019 (Action Plan) recommends specific actions, which if implemented, will substantially improve the health and well-being of Australians with CHD as well as the many family members and carers impacted by CHD.

This Evidence Compendium C provides the supporting evidence for the specific Recommendations of the Action Plan. The accompanying Consultation Compendium A describes the consultation process employed in the development of the Action Plan, including consultation with the clinical leadership of all state and territories paediatric and adult cardiology teams, as well as other non-government agencies and the broader CHD research community. Priorities and focus areas were developed under the direction of the expert CHD Action Plan Steering Committee and with guidance of the Australian Department of Health. Identification of priorities and key focus areas was informed by *The 2011 CHD White Paper*[1] and outcomes of the September 2017 HeartKids CHD Roundtable.

The following Action Plan priority areas and gaps were identified:

- Best practice models of care of all people living with and or impacted by CHD including infants, children, young people, adults and their families so that quality of life is improved and the burden of disease is reduced;
- Strategies to address the neurodevelopmental and mental health issues for patients and their families;
- Define the workforce and infrastructure priorities of CHD to ensure equity of services;
- Prioritise and identify gaps in CHD research to inform the Medical Research Future Fund which will now be distributed from the Mission for Cardiovascular Health 2019;
- Make recommendations on how to address the significant financial impact of CHD; and
- Develop the performance measures to better understand not just patient care but also tracking the implementation of the Action Plan itself.

The Action Plan priorities and focus areas included:

1. Management, care and support
   Focus Area 1: Standards of Care
   Focus Area 2: Infrastructure
2. Supportive Communities
   Focus Area 3: Awareness and Education
   Focus Area 4: Neurodevelopmental and Mental Healthcare
   Focus Area 5: Priority Populations
3. Research
   Focus Area 6: Research Priorities
   Focus Area 7: Surveillance

Linkages of priorities and focus areas with key policy documents

While there were significant linkages and associations between the priorities and recommended actions of this Action Plan with that of other relevant frameworks and plans, unique objectives and identified priorities and recommendations necessitate the development and implementation of this first Action Plan for CHD.

The identified Action Plan priority areas align with the Australian Government *National Strategic Framework for Chronic Conditions* (the Framework).[2] The Framework was approved by the Australian Health Ministers' Advisory Council in 2017 and outlines the key objectives and priority areas for Action Plans. The Action Plan's focus on standards of care and infrastructure aligns with the objectives of the Framework, with both aiming to provide 'efficient, effective and appropriate care to support people with chronic conditions'; to 'target priority populations'; and to provide services for improved diagnosis and intervention to reduce the burden of disease. The Action Plan also highlights the common challenges experienced by those with chronic conditions including the financial burdens and eligibility for NDIS, access to carers allowance, and eligibility and jurisdictional challenges of patient assisted travel schemes.

The benefits of early intervention and access to services are recognised in the Action Plan as well as the Framework, the *National Aboriginal and Torres Strait Islander Health Plan 2013-2023* and the *WHO Global Action Plan for Prevention and Control of Noncommunicable Diseases 2013-2020*. Significantly, the focus on the quality of life across the life course, beginning before birth, through infancy to adulthood, is a common objective shared by these plans.[3] [4] Common objectives include targeting priority populations, including young adults and challenges of transition, and identifying those at most risk. The importance of providing mental health patients and carers is critical. The challenges of rural, regional and indigenous communities in accessing support are significant. The Action Plan addresses CHD-specific issues which are also shared with those impacted with rheumatic heart disease (RHD).

While the Action Plan shares similar objectives with other plans, it can be distinguished by the need to address CHD-specific issues, including lack of standards of care, focus on childhood-specific care, neurodevelopmental co-morbidities, awareness and data, and workforce issues. Importantly, the lack of Australian standards of care is addressed in the Action Plan and represents an opportunity to develop not only the first Australian standards of care, but also the first international neurodevelopmental and mental health standards of care. Variability in paediatric models of care is not addressed in the other policy documents, and the opportunity for regularisation of clinical structuring could better facilitate delivery of outcomes identified in other plans.[5] [6]

The current lack of awareness of CHD among the general public, the CHD community, government agencies and health professionals poses a challenge to patients accessing support. Improved awareness and education may improve access to services and facilitate earlier interventions and better outcomes.[7] In addition, the causes of CHD remain largely unclear and are thought to result from a combination of genetic and environmental causes, necessitating further research and awareness and education.

Finally, a distinction of the Action Plan objectives is the recognition of the unique workforce challenge. There are significant deficiencies in specialist adult CHD services and workforce shortages, impacting patient outcomes,[8] and necessitating the recommendation to prepare the Australian health care system and community to provide a better and healthier future for those impacted by CHD.

CHD Prevalence and Impact

THE SIZE OF THE PROBLEM: PREVALENCE

Over 65,000 Australian children and adults live with CHD, and their many family members and carers are also impacted.[9] Congenital heart defects are the most common types of birth defects. Every day in Australia, eight babies are born with congenital heart disease; worldwide there are about 1.35 million babies with heart disease each year.[10] Congenital heart disease is one of the leading causes of death in Australian babies and of hospitalisations in the first year of life.[11] [12] [13]

In Australia, RHD is a highly significant form of CHD resulting from poorly diagnosed and treated rheumatic fever infections, and the vast majority of people with RHD are Aboriginal and Torres Strait Islander people, many of whom live in remote areas of central and northern Australia. Pacific Islanders and migrants from high-prevalence countries are also at high risk.[14]

For the first time in history, adults living with congenital and acquired CHD outnumber children with the disease, yet a lack of definitive research on the prevalence suggests the number of people living with CHD could be far higher than current estimates.[15] This includes around 32,000 children under 18 years of age, and over 32,000 adults who have lived with a heart condition since childhood. As Australian birth rates increase and enhanced medical care and technology continue to improve survival rates, the prevalence of congenital heart disease is expected to increase, particularly in the adult population where it is predicted to increase annually by five per cent.[16] [17]

Understanding the true prevalence of CHD in Australia is challenging. Prevalence data is often extrapolated from countries similar to Australia, using administrative databases, such as the Netherlands National CHD Registry, and the results are then applied to the Australian population.[18] [19] The most recent national data on the prevalence of congenital heart disease is from the 2003 Australian Congenital Anomalies Monitoring System. The recently-launched CHAANZ Congenital Heart Disease Registry will be critical in providing essential information on CHD in Australia (CHAANZ). Data about the prevalence of CHD in Australia, as well as the burden of disease, is needed to provide an evidence-based platform for setting priorities and guiding health policy and strategies.

Mortality

Congenital heart disease is a leading cause of death of Australian babies. Among infants, perinatal and congenital conditions caused most deaths (77 per cent of deaths).[20] Historically, survival of congenital heart disease patients with significant lesions beyond early childhood was limited, with less than one-third of infants being expected to survive into adulthood. The number of deaths from CHD in Australia has significantly declined over the last three decades, particularly in children. Most (more than 95 per cent) of people with CHD now survive into;[21] [22] however, life expectancy continues to be below normal.[23] [24]

Inequalities in outcomes for young Aboriginal and Torres Strait Islander people with RHD are particularly pronounced. Young Indigenous people (younger than 35 years) are 122 times more likely to have RHD than their non-Indigenous peers.[25] Between 2011–2015, 94 per cent of people diagnosed with acute rheumatic fever identified as Indigenous and 55 per cent of them were aged zero to 14 years.[26] On average, Aboriginal people with RHD die at 41 years of age.[27]

Morbidity of CHD

Despite remarkable medical success in recent years, CHD continues to be a major global health burden.[28] People with CHD and their families have a poorer quality of life than the general

population and suffer higher rates of mental health problems including depression, anxiety and Post Traumatic Stress Disorder. There is clear evidence that people of all ages with congenital heart disease are at greater risk of lower health-related quality of life compared with their healthy peers, particularly in relation to neurodevelopmental, psychosocial and behavioural outcomes.[29] [30] [31] [32] [33] [34] Families of people with CHD, including parents, carers and siblings, also experience significant hardship and suffering.[35]

Psychological morbidity

Although medical advances have reduced paediatric mortality and extended life expectancy for those with CHD, psychological morbidity remains prevalent. Reported prevalence of anxiety is higher than depression, both of which have been generally found to exceed population norms, ranging from 15 to 33 per cent in patients of all ages.[36] [37] [38] [39] [40] Our understanding of the psychology of adults with CHD lags decades behind our knowledge of children's experiences. Some research has found that one in three adults with CHD report symptoms of anxiety or depression warranting intervention.[41] The vast majority of these adults go untreated.[42]

The presence of anxiety and depression is associated with a range of difficulties, including but not limited to increased risk of major cardiovascular events, shorter event-free survival,[43] lower adherence to cardiac care recommendations,[44] and impaired health- related quality of life (HRQOL).[45] Higher levels of cardiac events can be a major burden on Australia's health care system and levels of workforce participation and productivity.

Correlates of anxiety and depression include lower perceived health status,[46] the presence of neurodevelopmental delay,[47] greater social isolation,[48] and poorer self-perception.[49] Approximately 30 to 80 per cent of parents of children with complex congenital heart disease also reports high psychological distress.[50] Poorer parental mental health is associated with greater suicidal ideation, poorer physical health and greater healthcare usage, in addition to poorer family functioning and child emotional, cognitive, behavioural, social and health-related outcomes.[51]

Families of people with CHD including parents and carers experience significant hardship and suffering.[52] [53] Parents of children with CHD experience more anger and sadness than parents of healthy children,[54] more social problems and fewer leisure activities,[55] greater feelings of distress and hopelessness,[56] and higher levels of overall stress than parents with children with other chronic conditions.[57] [58] Research shows that one in three mothers and fathers of infants with complex CHD report symptoms that meet clinical criteria for depression, and about 50 per cent of parents report severe stress reactions consistent with a need for clinical care up to one year after their baby's diagnosis.[59] These rates far exceed documented rates of perinatal depression and anxiety in the general community[60] and healthcare providers often markedly underestimate the severity and potential consequences of these symptoms.[61]

Morbidity associated with the impact of surgery

Children who undergo open heart surgery, particularly in the first year of life, are known to have a higher risk of impairment across multiple domains when compared with the general population including intellectual ability, behaviour, social interaction, communication and executive function[62] [63] The underlying cause of these deficits is multifactorial, with complex interactions between genetic susceptibility, physiological consequences of the heart defect and circulation, perioperative brain injury and psychosocial factors all contributing.[64] [65]

Children with complex congenital heart disease (including those who need surgical intervention in the first year of life and those with other, co-morbid physical conditions) are at increased risk of

neurodevelopmental impairment and disability.[66] The risk and severity of neurological impairment increase with greater CHD complexity, the presence of a genetic disorder or syndrome, and greater child and family psychological stress.[67] During infancy, the most pronounced difficulties occur in motor functioning. By early childhood, neurological impairment is characterised by difficulties in fine and gross motor skills, speech and language, attention, executive functioning, emotion regulation and behaviour.[68]

Studies also show that children with complex CHD are also up to four times more likely to be diagnosed with attention deficit and hyperactivity disorder (ADHD) than the general population.[69] People with CHD have a greater risk of neurodevelopmental impairment and disability including developmental delay and other learning difficulties. The neurodevelopmental and psychosocial morbidity related to CHD and its treatment can have a lasting negative impact on educational achievement, employability, lifelong earning potential and quality of life for many survivors, creating a burden of disease on the individual, family and society.

The Burdens of CHD and the Need for the CHD Action Plan

The economic burden of CHD has not been well described, and it is difficult to estimate the total economic impact of CHD on the healthcare and education systems, governments, workforce and people affected by CHD. However, it is evident that CHD poses a substantial economic burden on Australia's health care system. Congenital heart disease is one of the leading causes of hospitalisation in the first year of life,[70] and successful treatment requires highly specialised care. CHD requires extensive financial resources both in and out of the hospital, with costs directly related to the degree of disease complexity. A Hong Kong study calculated the annual cost of management of simple ACHD was USD 2,638 and complex ACHD was USD 6,425.[71]

Surgery for CHD results in an enormous financial burden for most families at a very vulnerable time.[72] Significant out-of-pocket costs exist for patients and their families, including travel and accommodation for families required to travel to the designated CHD specialist centres. This burden is particularly high for families in rural and remote locations including Aboriginal and Torres Strait Islander families.

Social burdens of CHD including education, employment and insurability, which increase the societal costs of adult CHD, are now being recognised for adults living with CHD and financial stress experienced by patients and their families and carers can be significant.[73] A recent study showed that over 40 per cent of people with CHD have to travel farther than 200 kilometres for specialist treatment, and have significant out-of-pocket expenses of AUD 2,500-AUD 3,500.[74]

The large and growing population of adults with CHD requires lifelong and highly- specialised medical care. This changing demographic and epidemiology of CHD have 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. These emerging challenges place pressure on Australia's healthcare system, highlighting gaps in service delivery and family support. Additionally, due to a lack of coordinated national effort to date, CHD is characterised by:

- a lack of data
- the absence of national standards of care
- deficiencies in specialist adult CHD services
- workforce shortages.

This strong body of evidence highlights the significant impact of CHD on the many thousands of patients and their families and carers, and provides the foundation for all recommendations of the

Action Plan, reinforcing the call for urgent action to provide holistic person-centred and family-centred care for CHD across the life course.

Priority 1 Management, care and support

Standards of Care

Despite excellence in many areas of cardiac care, Australia has never had nationally agreed standards of care for CHD. Significantly, there is only limited and, in some cases, no significant mention of CHD or its management in state based cardiac services frameworks or general clinical frameworks listed below:

- Victorian Cardiology Framework[75]
- Queensland Aboriginal and Torres Strait Islander cardiac health strategy 2014-2017[76]
- Queensland Government Department of Health Cardiac Services[77]
- South Australia Clinical Services Capability Framework[78]
- Tasmanian Role Delineation Framework[79]
- Cardiac Clinical Advisory Group Cardiology Services[80]
- Cardiac Rehabilitation and Secondary Prevention: A framework for the Northern Territory 2012[81]
- WA Health Clinical Services Framework 2014-2024[82]
- National Strategic Framework for Chronic Conditions[83]
- New Directions for Cardiac Care in the NT 2017[84]

Securing quality care across the life course can be challenging and certain stages require urgent attention, such as transition of care from paediatric to adult cardiac health services, where over 50 per cent of Australians with CHD are being 'lost to care'.[85] [86]

Without standards of care, risks to patients include: inconsistency of practice and resourcing of patient care, particularly disadvantaging people with CHD in rural, remote and isolated communities; fragmented care and focused on 'moments of care' rather than lifelong 'continuity of care'; lack of alignment between workforce and infrastructure planning and resourcing; and CHD patients being 'lost to care',[87] [88] and suffering associated consequences for the appropriate management of their disease, mental health, well-being and life expectancy.[89] Importantly proactive development of standards of care can avoid the need to address adverse events which have previously, at major clinical centres in the UK, resulted in the Bristol Enquiry and the need to develop standards of care.[90]

International examples of standards of care include:

- National Health Service England Congenital Heart Disease Standards and Specifications[91]
- Specialist Congenital Heart Disease standards for children and adults in Scotland[92]
- 2018 AHA/ACC Guideline for the Management of Adults with Congenital Heart Disease (August 2018) (USA)[93]
- Adult Congenital Heart Association Strategy (USA)[94]

Two major guidelines, 2018 AHA/ACC Guideline for the management of Adults with Congenital Heart Disease (August 2018) (USA) and the Adult Congenital Heart Association Strategy (USA) highlight the absence of standards for childhood care.

Box 1 provides an overview of the categories contained in the Specialist Congenital Heart Disease standards for children and adults in Scotland. These standards highlight the whole patient pathway and address many of the previously identified risks to patients in Australia.

BOX 1: CATAGORIES OF THE SCOTTISH STANDARDS OF CARE		
The Scottish standards encompass the whole patient pathway and are subdivided into categories:		
A	-	The Service Approach
B	-	Staffing and Skills
C	-	Facilities
D	-	Interdependencies
E	-	Training and Education
F	-	Organisation, Governance and Audit
G	-	Research
H	-	Communication with Patients
I	-	Transition
J	-	Pregnancy and Contraception
K	-	Fetal Diagnosis (paediatrics only)
L	-	Palliative Care and Bereavement
M	-	Dental (Scotland, 2018).

Australian statements and guidelines include:

- CSANZ 2013 Adult CHD Recommendations for Standards of Care Position Statement[95]
- CSANZ 2016 Adult CHD Recommendations for Standards of Care Position Statement[96]
- Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease[97]
- ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease: Executive Summary[98]

The available international standards of care and current Australian guidelines will inform the recommended actions to provide best practice standards of care and guidelines:

- *Recommendation 1.1 Develop national standards of care for CHD*
- *Recommendation 1.2 Develop authoritative national clinical practice guidelines on CHD for health services and health practitioners throughout Australia*
- *Recommendation 1.3 Establish a CHD taskforce to oversee the development of standards of care for CHD plans.*

Standards of care for CHD will be tailored to the unique Australian context and will enhance existing cardiac services provided across the country. The standards will include components that build on existing international and Australian resources including care of adults with congenital heart disease [the Adult Congenital Heart Disease Recommendations for Standards of Care developed by the Cardiac Society of Australia and New Zealand[99] [100]] and care for priority populations through the service standards for equitable cardiovascular care for Aboriginal and Torres Strait Islander People.[101]

A number of CHD clinical practice guidelines exist, such as the Australian guideline for prevention, diagnosis and management of acute rheumatic fever and RHD;[102] however, significant gaps exist. Following the development of standards, models of care, care pathways and clinical practice

guidelines will be developed as required, alongside other bodies of work that may be identified during the development of the standards and in response to areas of need, such as gaps between the care defined by the standards and current practice.

A priority area in *Recommendation 1.2* is the development of guidelines for echocardiographic screening for RHD. Relevant guidelines for echocardiographic screening for RHD in indigenous Australian children would enable earlier diagnosis of RHD and be informed by the previous review of World Heart Federation echocardiographic criteria for RHD.[103]

The CHD Action Plan Steering Committee and participants contributing to this CHD Action Plan highlighted that the lack of awareness of CHD, and the lack of recognition of CHD or its management in the previous state based cardiac services frameworks or general clinical frameworks and plans, represented a significant challenge for patient care.

The establishment of a CHD taskforce to oversee the development of standards of care for CHD plans was proposed to ensure that standards and of care and clinical practice guidelines were developed in accordance with NHMRC guideline standards, regularly reviewed for currency and relevance, and that workforce and infrastructure plans were aligned to the standards of care.

Infrastructure and Workforce

Australia remains under-resourced and inconsistent with international guidelines, with workforce shortages and skill gaps limiting access and compromising patient quality of care.[104] Workforce shortages are present across paediatric and adult services, although it is particularly problematic for the adult CHD (ACHD) workforce due to the large and growing cohort of adults with CHD.

As a result, many adults with CHD have been lost to follow-up. Recent data noted an alarming rate of 'loss to follow-up' for people with CHD of between 50 to 75 per cent, which can have catastrophic health impacts.[105] [106] [107] [108] Common reasons for 'loss to follow-up' include: insufficient patient/carer preparation, inadequate service integration and limited access to adult CHD specialists;[109] [110] and being cared for by general cardiologists without specialist training result in a high risk for adverse clinical outcomes.[111] [112]

A critical infrastructure issue is the shortage of dedicated facilities for CHD. This is particularly challenging for people living in regional, rural and remote areas due to Australia's vast geography. Australia faces unique geographical challenges when delivering accessible healthcare. The challenges of geographical spread, low population density and limited infrastructure, as well as the higher costs of delivering rural and remote healthcare, can affect access to healthcare.[113] Greater co-location of family support services within hospital settings would help to address this.

The CHD Action Plan Recommendations aim to improve the capacity of the workforce and ensure the required infrastructure to provide quality care, improving planning, training and education, support of primary care providers and sharing information using approaches that can interact throughout Australia, *Recommendation 2.1 Develop a CHD infrastructure and workforce plan* to addresses the need to plan for future required capacity and capability based on best practice.

Such a CHD infrastructure and workforce plan would be informed by international guidelines and specific Australian needs. In 2010, there were approximately 30 paediatric cardiologists in Australia[114] yet the equivalent of only six full time cardiologists care for adults with CHD.[115] [116] [117] [118] This equates to one FTE ACHD specialist per 3.5 million population in Australia, in contrast to international recommendations of one FTE ACHD specialist per one million population or one ACHD centre per two million population.[119] [120]

The recent NHS UK review of the ACHD services recommends at least four FTE congenital cardiologists and at least four FTE congenital surgeons for a Level 1 ACHD specialist centre (this is similar to a comprehensive adult congenital heart centre (CACH) in Australia).[121] Specific recommendations are also for Level 2 and 3 ACHD Centres (these are equivalent to the regional adult congenital heart centres (RACH) in Australia). Based on international best practice of one centre for every two million population,[122] Australia should have at least 12.5 CACH centres based on a population of 25 million; yet, there are only 5 designated CACH centres across Australia and no RACH centres.

More recently, the Adult Congenital Heart Disease Working Group of the Paediatric and Congenital Council and CHAANZ) has recommended:[123]

- Comprehensive ACH centres, servicing regions with a population >2–3 million, should be staffed with least two ACHD specialists, ACHD surgeons, a clinical nurse co-ordinator, a nurse educator, and sonographers with congenital cardiac expertise, with appropriate access to clinical psychology, genetic counselling and social work.
- Regional ACH centres should be staffed by at least one ACHD cardiologist, a nurse co-ordinator and appropriately trained cardiac sonographers with links to a comprehensive ACH centre.

Recommendation 2.2 Invest in technological infrastructure is supported by evidence that such investment benefits patients and the health system. During development of the CHD Action Plan, no evidence was clearly identified of the adaptation or uptake of e-health technologies to reduce the needs of rural and remote patients to travel (i.e. video conference). eHealth educational interventions designed for people with low eHealth literacy can increase eHealth literacy and confidence, irrespective of age, ethnicity, education, and previous internet use and eHealth is highly desired by parents of children with complex CHD;[124] [125] such technological investment would be applicable.

Investment in technological infrastructure and development of education and training to support health professionals and disseminate best practice information also address *Recommendations 2.3, 2.4 and 2.5* and improve the capabilities of both workforce and infrastructure to optimise resources.

Priority 2 Supportive communities

Awareness and Education

The lack of investment and overall infrastructure in CHD (beyond surgical interventions and clinical heart care) leaves many families isolated, with no clear pathway of care to seek information and support for suspected neurodevelopmental delay, behavioural and mental health issues for their child.[126]

Although information and support are currently available, these may not be evidence-based, appropriately targeted or resourced. The lived experience of families supports the research that shows that parental information on CHD is insufficient and suggests that there is a link between information dissemination and satisfaction with care.[127] Parental information on CHD is particularly lacking for priority populations with unique cultural needs, such as Aboriginal and Torres Strait Islander families where differences in health literacy and English literacy may compound communication challenges.[128] [129] Also, young people with CHD generally possess poor knowledge of their condition nor awareness of the need for longer, follow-up care.[130] [131]

A study commissioned by HeartKids in 2018 highlighted that health professionals have limited knowledge or awareness of CHD.[132] The research also showed that barely a quarter of people surveyed are aware of HeartKids (as the only peak consumer organisation focused on supporting people impacted by CHD), congenital heart disease and even less aware of its impact on individuals and families.[133]

Education for families and patients, and the community more broadly, including community settings, such as schools, work places, financial institutions and sports and recreation groups, can ensure information about support and required intervention is available to assist people to navigate the system[134] and such education has been shown to improve the understanding of CHD, particularly in young adults with CHD.[135]

The effective provision of such information and education is proposed in *Recommendation 3.1 Increase access to quality, relevant information, education and support through a range of approaches including phone, face to face and digital*. In addition to developing evidenced-based quality information and education, the methods of engagement and delivery should be broad and appropriate for the target population to improve accessibility and accommodate a range of health literacy needs and learning styles. For instance, delivery of text messages to young adults has been shown to improve engagement with the transition service for young adults in renal clinical care, a model that can be adopted for support of young adults in transition from paediatric to adult cardiac health services.[136] The development of eHealth educational interventions to increase eHealth literacy and confidence has been shown to be relevant for patients regardless of irrespective of age, ethnicity, education, and previous internet use and eHealth is highly desired by parents of children with complex CHD.[137] [138] Engaging non-health professionals to create supportive communities for those with CHD is also proposed in *Recommendation 3.4 Deliver awareness and education for non-health professionals*.

Peer support programs, such as those provided by HeartKids, are recognised as being beneficial and effective for patients with chronic and complex conditions.[139] [140] Peer support programs designed specifically for those with CHD have been shown to significantly improve the psychological wellbeing, alleviate feelings of loneliness, and empower people impacted by CHD.[141] Such peer support programs were shown to be even more effective for some when integrated with clinical psychology services.[142] Peer-to-peer support provided by 'veteran' neonatal intensive care unit

(NICU) to those with current NICO babies have also been shown to be effective.[143] HeartKids has recently supported and delivered the pilot version of the HeartKids Family Coping Program for families impacted by CHD, and results indicated that parents who completed the program had increased coping self-efficacy even 6 months after the completion of the course.[144] The value of peer support is only realised if support team members can deliver more and expanded programs and this is reflected in *Recommendation 3.2 Increase access to ongoing support by funding inpatient and outpatient community (peer) support team members.*

Components of *Recommendation 3.3 Increase the number of young people engaged with transition and ongoing specialist CHD care* are also addressed in the evidence for proposing *Recommendation 5.3 Establish models of care for the transition of care from paediatric to adult cardiac health services*. Development and expansion of relevant delivery of information education and peer support programs can be applied to engagement and retain young adults with the health service. Best practice models of peer support in an integrated paediatric to adult clinical service model for young adults with kidney disease can be adopted to engage young people with CHD.[145]

Neurodevelopmental and Mental Healthcare

Survival for children with CHD has significantly increased with most surviving into adulthood, yet CHD remains a global health burden and has a profound impact on the health and wellbeing of those affected, with far-reaching effects that ripple through a family and across a lifetime.[146]

People of all ages with CHD are at greater risk of lower health-related quality of life compared with their healthy peers, particularly in relation to neurodevelopmental, developmental delay and other learning difficulties, psychosocial and behavioural outcomes. People with CHD as well as their families have a poorer quality of life than the general population and suffer higher rates of mental health problems including depression, anxiety and post-traumatic stress disorder (PTSD).[147] [148] [149] [150] [151] [152]

When considering the experiences of those impacted by CHD, parents of children with CHD experience challenges practising positive coping strategies, and coping-focused psychological support throughout the parents' and children's journey may be of benefit.[153] Significantly, fear of death and the psychological well-being of their children were the main parental concerns, while in adults with the Fontan correction for the congenital form of CHD, fear of death was the main concern, and may be addressed by providing family-focused psychological interventions to improve the psychosocial functioning of both parents and young people.[154] [155] [156]

Optimal care for CHD involves cardiac care that integrates neurodevelopmental and mental healthcare as core business.[157] [158] An emphasis on earlier interventions and quality of life across the life course, beginning before birth, through infancy to adulthood, is required.[159] Early intervention can make a profound psychological difference for families, promoting maternal adjustment and development in infants.[160]

Available mental health plans provide support for common issues experienced by those with chronic conditions, however, evidence on the efficacy of proven mental health interventions which have been tested in a range of other settings is severely lacking in CHD. There is no specific mention of CHD in the World Health Organisation Mental Health Action Plan 2013-2020.[161] The Fifth National Mental Health and Suicide Prevention Plan,[162] endorsed by the Council of Australian Governments Health Council (COAG Health Council), does not specifically address evidenced-based interventions addressing the impacts of CHD, but targets eight priority areas, which reinforce the particular needs of priority populations, including:

- Achieving integrated regional planning and service delivery
- Effective suicide prevention
- Coordinated treatment and supports for people with severe and complex mental illness
- Improving Aboriginal and Torres Strait Islander mental health and suicide prevention
- Improving the physical health of people living with mental illness and reducing early mortality
- Reducing stigma and discrimination
- Making safety and quality central to mental health service delivery
- Ensuring that the enablers of effective system performance and system improvement are in place.

The Fifth National Mental Health and Suicide Prevention Plan does not provide significant guidance in relation to the impact of critical or chronic disease on mental health. While it does cover the impact of mental illness on physical health, it does not provide guidance on the impact of physical illness on mental health. In addition, the Fifth National Mental Health and Suicide Prevention Plan does not directly address three areas of profound significance in CHD:

- Neurodevelopmental disability and neurodevelopmental care
- Perinatal mental health, particularly mental health in pregnancy and the impact of this for the developing fetus and child
- Integration on mental health (including neurodevelopmental) services across the spectrum of paediatric services, including hospitals, community-based health services, and child care and schools.

Recommendation 4.1 Develop standards of mental healthcare for CHD is based on available plans as well as evidence of psychological interventions shown to be effective for people affected by childhood-onset heart disease[163] and the need for routine screening of health-related quality of life (HRQOL) for families of young children with complex CHD to identify those predictors of HRQOL that are modifiable through prevention and intervention.[164]

The significant need to recognise and address neurodevelopmental, developmental delays, and other learning difficulties will be supported by recent research evidence of best practice[165] and *Recommendation 4.2 Develop standards of neurodevelopmental care for CHD*, and *Recommendation 4.3 Establish clear models of care.* Implementation of these recommendations is likely to have implications for critical and chronic illness well beyond heart disease.

Priority Populations

CHD impacts all Australians, but some priority populations are disproportionately affected due to a complex interaction between the physical environment, social and cultural determinants and bio-medical and behavioural risk factors.[166] This is demonstrated by a higher prevalence of CHD and a greater burden of disease, resulting in inequitable health outcomes.[167]

Barriers to effective treatment among these priority populations include remoteness,[168] [169] [170] lack of culturally appropriate healthcare services, legal and institutional barriers to accessing support, individualised and holistic transition support, care and coordination required for adolescents and young adults and their parents/carers at the critical transition stage in the cardiac care pathway.[171] [172]

Around seven million Australians, about 29 per cent of the population, live in rural and remote areas.[173] These people face unique challenges due to their geographic isolation, and they often have poorer health outcomes than people living in major cities.[174] Over 40 percent of people with CHD

have to travel over 200 kilometres for specialist treatment and have significant out-of-pocket expenses of AUD2,500-3,500.[175] These poorer health outcomes may be due to factors such as disadvantage in education, employment opportunities, income and access to services.[176]

Both congenital forms of CHD and RHD are more common among Aboriginal and Torres Strait Islander people compared with non-Indigenous Australians, and Indigenous Australians experience worse outcomes than non-Indigenous Australians.[177]

The National Aboriginal and Torres Strait Islander Health Plan 2013-2023 identifies the health needs of Aboriginal and Torres Strait Islander People and strategies to address identified needs.[178] The Commonwealth Government has recognised the need to close the gap between the health outcomes of Aboriginal and Torres Strait Islander people and the broader population.[179] Cardiac measures for Aboriginal and Torres Strait Islander people have been highlighted in national and international publications.[180] [181]

The importance of empowerment of Aboriginal and Torres Strait Island People in determining health priorities and outcomes has been emphasised in publications.[182] And *Recommendation 5.1 Fund CHD cardiac coordinators in each jurisdiction* is proposed to engage the community in a culturally sensitive and appropriate model that can service regional and rural areas, thus also addressing *Recommendation 5.5 Investigate options for reducing barriers to accessing care and supporting people with CHD and their families*. Care coordination may be delivered with enhanced outreach services in *Recommendation 5.3 Evaluate current specialist outreach services for regional and remote communities, including remote Aboriginal communities, and fund outreach services where significant gaps exist*.

Recommendation 5.2 Fund cardiac sonographers and a portable echo machine in each jurisdiction is based on relevant guidelines for echocardiographic screening for RHD in indigenous Australian children and would enable earlier diagnosis of RHD; such an approach has been shown to be a cost-effective model appropriate for expanded application to improve clinical and economic outcomes.[183]

The significant challenge of providing individualised and holistic models of transition of care from paediatric to adult cardiac health services is addressed in *Recommendation 5.4 Establish models of care for the transition of care from paediatric to adult cardiac health services, including investing in transition nurses*. Evidence from patient and family consumer research has identified that transition programs should be dedicated, have youth-friendly and holistic focus (in particular around emotional wellbeing), and include parental involvement and coordination efforts between paediatric and adult care with strong integration and prioritisation in the health system.[184] [185] [186] The adoption of relevant models shown to be successful for other chronic conditions, such as an integrated paediatric to adult clinical service in the UK for young adults with kidney failure, could be assessed for applicability to young adults with CHD.[187] These models are relevant to vulnerable populations including young adults at the critical stage of transition as well as the indigenous CHD community.

Priority 3 Research

Research Priorities

During the consultation process for development of this National Strategic CHD Action Plan, the Expert Working Group for Research considered a number of areas of research and identified key priorities that they determined would have the most impact on those impacted by CHD.

Experts reviewed the current needs of the patients and families impacted by CHD, informed by the topics discussed in other Expert Working Groups and from their own professional clinical and research experience, and highlighted the current health, social and economic burdens of CHD.

The Group first considered the main issues, including:

- No COAG/NHMRC/MRFF agreed Congenital or Acquired Heart Disease research framework or strategy exists.
- Australian CHD research investment has not kept pace with international investment.
- Clinical trials leading to new treatments have stalled in recent years
- Pharmaceutical investment in CHD research has been declining as companies favour large patient populations countries.
- CHD specialists have heavy clinical workloads reducing their capacity to participate fully in research.
- Major research units and thus investment are centred in Sydney and Melbourne. There is little investment in growing research centres in other states and territories.

The Group then considered the following questions:

- What will advance Australia's leadership, capability and capacity in CHD research?
- What are the identified CHD research priorities in the following categories which can best address the current needs of those impacted by CHD:
    - Childhood origins of congenital and acquired heart disease
    - Genomics
    - Cardiac regenerative medicine
    - Devices including conduits and patches
    - Clinical and research registries
    - Clinical trials and translation
    - Neurodevelopment
    - Mental health and resilience
    - Indigenous care
    - While of life care and transition of care
    - Genetic counselling
    - Exercise physiology and rehabilitation
    - Health economics?

While valuable research questions were discussed in each research category, the identified research priorities described in the Action Plan were selected for their greatest potential impact, were consistent with those identified internationally,[188] and leveraged the significant collaboration opportunities within the Australian CHD patient and clinician and research communities.

Research priorities identified:

- *Research Priority 6.1 Understanding the genetic causes of and predisposition to CHD to provide support for those impacted by CHD*

- *Research Priority 6.2 Preventing neurodevelopmental and mental health complication*
- *Research Priority 6.3 Understanding the impact of prenatal and postnatal factors affecting the health of children born with CHD and their families*
- *Research Priority 6.4 Undertaking longitudinal impact and informed practice with the National Congenital Heart Disease Registry*
- *Research Priority 6.5 Exploring the role of exercise and increased activity in reducing the impact of CHD.*

The Expert Working Group in Research also highlighted the need to invest the research funds strategically, acknowledging that whilst there are many worthwhile projects, there are just a few key projects within each nominated research priority that need to be initiated to realise synergies between research priorities, further build the research capability and CHD workforce in Australia. For this reason, it was proposed that calls for projects should be well targeted rather than general in nature, and address the key questions within the research priorities.

Research priorities 6.1, 6.2 and 6.3 aimed to address the greatest needs of those with CHD. While progress has been made in understanding the complex genetic and environmental contributors to CHD,[189] a priority was understanding the causes of CHD as well as and opportunities for prevention. The significant co-morbidities discussed in this Compendium and in the Action Plan necessitated a focus on developing and accessing early interventions to improve neurodevelopmental, developmental and mental health. The benefits of being able to identify individual characteristics both before and after birth that influence long-term outcomes for both child and family are also a priority. Priorities also reflected an appreciation that interventions may be required across the life course for patients as well as family members.[190]

A key approach identified by researchers was the prospective acquisition of clinical, social and genetic information from a cohort of children born with CHD. This would provide access for investigators from multiple disciplines and significantly facilitate a number of research priorities. Such a cohort, followed indefinitely, would be of international significance. The equivalent in the long-term, ongoing adult cardiovascular cohort study, the Framingham Study in New England examining risk factors for coronary artery disease.[191] The other advantage to the initiation of such a study is that it involves all clinical services across Australia – it will provide insights not just relevant to research priorities but also health services design by highlighting areas of practice variation across jurisdictions. Researchers in Australia have demonstrated their ability to work together in initiatives such as the Fontan Registry and have already come together to endorse a neonatal cohort study entitled 'Growing up in Australia with CHD'.

Children born with CHD often experience reduced exercise capacity, and those with severe CHD in the past were not even encouraged to be active or exercise. Exercise training can improve exercise capacity and quality of life for adults with acquired and congenital heart disease.[192] [193] [194] [195] Resistance training can improve cardiac output and exercise capacity in those patients with severe congenital heart conditions and correction with the Fontan.[196] However, some studies to date suffer from suboptimal trial designs, low patient numbers, and homogeneity of investigated cardiac anomalies, additional research and trials are required to identify the types of exercise, and their specific benefits.[197]

While exercise and activity can be a benefit for those with CHD, long-term commitment to regular physical activity can be difficult to maintain in patients. Additional lifestyle changes through behaviour modification and individualised counselling may also provide improved health outcomes, necessitating further research on the impacts of specific interventions in exercise, physical activity

and diet and nutrition.[198] The striking thing about research in this field is that exercise in patients with single ventricles may be more effective than any pharmacological intervention in terms of improving cardiac performance.[199] Such gains may defer or prevent the onset of symptoms and medical care for many CHD patients.

The National CHD Registry is key to achieving the research priorities and undertaking a longitudinal impact to inform practice. The CHD clinical community has recognised the need for a lifetime continuum of care to achieve the best health outcomes.[200] [201] The current Fontan Registry demonstrated that such CHD registries are useful for tracking the impact of interventions, informing changes of clinical practice, supporting research questions to identify prevalence and mortality, and for piloting the benefits of patient engagement to improve patient access to information, support and improved health and wellbeing.[202] More recently, the Australian CHD clinical and research community highlighted and developed the first bi-national registry of all those with CHD (National CHD Registry), to be able to determine the impact of interventions to improve future care.[203]

Research Surveillance

A number of surveillance activities and research efforts are proposed in this Action Plan to address the needs of those impacted by CHD and to inform the future needs of the healthcare system. Surveillance is defined as an ongoing, systematic collection, analysis and interpretation of health-related data essential to the planning, implementation, and evaluation of public health practice.[204] Surveillance recommendations include:

- *Recommendation 7.1 Invest in the Congenital Heart Alliance of Australia and New Zealand (CHAANZ) Registry*
- *Recommendation 7.2 Invest in an economic burden of CHD report*
- *Recommendation 7.3 Invest in funding of the annual national CHD survey*
- *Recommendation 7.4 Develop an evaluation framework that includes the production of an annual report card to monitor progress against this Action Plan.*

The Congenital Heart Alliance of Australia and New Zealand (CHAANZ) is developing the Australia and New Zealand Congenital Heart Disease Registry which will inform the future needs of the CHD patients and the healthcare system.[205] CHAANZ has also developed and distributed the National CHD Survey to those impacted by CHD (Nat CHD Survey description) with the results helping to inform the self-reported burden of the disease.[206] As well, CHAANZ and the National CHD Survey will inform an annual report card to monitor progress in support for those impacted by CHD. Accelerated implementation of the pilot CHAANZ National Congenital Heart Disease Registry (and harmonisation of several existing smaller condition-specific registries) will lead to: an accurate measure of the burden of CHD on the Australian community; an evidence-based needs assessment tool; a means of re-engaging those in danger from being lost to follow-up care; and a resource for the appropriate recruitment of subjects for clinical trials.

The Australian and New Zealand Fontan Registry (Fontan Registry) has developed significant patient engagement and research which informs the specific needs of patients and the healthcare system.[207] The interactions of the Fontan Registry with patients and their families within Australia and New Zealand is an established model for the involvement of patients and parents that may result in research that is more relevant, focussed, and practically applicable in a healthcare setting.[208] Evidence reviewed in Stokes and Oliver 2017, indicates that such community engagement impacts positively on various health outcomes across a variety of conditions.[209]

The CHD community, the healthcare system, the workforce and the broader economy would benefit from more clarity of the economic burden of CHD to inform future planning.[210]

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