



# NATIONAL STRATEGIC ACTION PLAN FOR CHILDHOOD HEART DISEASE

February 2019

**BEYOND THE HEART : TRANSFORMING CARE**



**Australian Government**

**Department of Health**

©Commonwealth of Australia as represented by the Department of Health 2019

### Creative Commons Licence



This publication is licensed under the Creative Commons Attribution 4.0 International Public License available from <https://creativecommons.org/licenses/by/4.0/legalcode> (“Licence”). You must read and understand the Licence before using any material from this publication.

### Restrictions

The Licence may not give you all the permissions necessary for your intended use. For example, other rights (such as publicity, privacy and moral rights) may limit how you use the material found in this publication.

The Licence does not cover, and there is no permission given for, use of any of the following material found in this publication:

- The Commonwealth Coat of Arms. (by way of information, the terms under which the Coat of Arms may be used can be found on the Department of Prime Minister and Cabinet website <http://www.dpmc.gov.au/government/commonwealth-coat-arms>);
- any logos and trademarks;
- any photographs and images;
- any signatures; and
- any material belonging to third parties.

### Attribution

Without limiting your obligations under the Licence, the Department of Health requests that you attribute this publication in your work. Any reasonable form of words may be used provided that you:

- include a reference to this publication and where, practicable, the relevant page numbers;
- make it clear that you have permission to use the material under the Creative Commons Attribution 4.0 International Public License;
- make it clear whether or not you have changed the material used from this publication;
- include a copyright notice in relation to the material used. In the case of no change to the material, the words “© Commonwealth of Australia (Department of Health) 2019” may be used. In the case where the material has been changed or adapted, the words: “Based on Commonwealth of Australia (Department of Health) material” may be used; and
- do not suggest that the Department of Health endorses you or your use of the material.

### Enquiries

Enquiries regarding any other use of this publication should be addressed to the Branch Manager, Communication Branch, Department of Health, GPO Box 9848, Canberra ACT 2601, or via e-mail to [copyright@health.gov.au](mailto:copyright@health.gov.au)

***“Transforming childhood heart disease care in Australia is the best possible way for us to support people with childhood heart disease and their families to live longer, healthier and more productive lives.”***

Jan McClelland AM,  
Chair, HeartKids

***“To the childhood heart disease pioneers, the first generation of survivors, families and medical staff, this Action Plan is the embodiment of decades of love, loss and hope. It has lit the path that many of us forged for future generations. Thank you.”***

Elle,  
adult living with CHD

We acknowledge Traditional Owners across this land and recognise the strength, resilience and capacity of First Nations Peoples. We pay respect to Elders past, present and emerging.

Development of the National Strategic Action Plan for Childhood Heart Disease February 2019 was led by HeartKids Ltd with funding from the Australian Government Department of Health.





# TABLE OF CONTENTS

- EXECUTIVE SUMMARY ..... 5
- ACRONYMS ..... 7
- CHILDHOOD HEART DISEASE (CHD) AND THE EMERGING CHALLENGES..... 8
- THE GOAL AND THE APPROACH ..... 12
- PRIORITY 1: MANAGEMENT, CARE AND SUPPORT ..... 14
- PRIORITY 2: SUPPORTIVE COMMUNITIES..... 21
- PRIORITY 3: RESEARCH..... 31
- ACKNOWLEDGEMENTS ..... 40
- APPENDIX 1: FRAMEWORK FOR ACTION ..... 41
- APPENDIX 2: GLOSSARY ..... 44
- REFERENCES ..... 48



## EXECUTIVE SUMMARY

The National Strategic Action Plan for Childhood Heart Disease 2019 - Beyond the Heart: Transforming Care (the Action Plan) aims to reduce the impact of childhood heart disease (CHD) in Australia and achieve the goal that people with CHD live longer, healthier and more productive lives through effective management of CHD across the life course.

CHD, which includes congenital heart disease and acquired heart disease, places a significant burden on the estimated 65,000 children and adults living with CHD ('heart kids') and their family members and carers.<sup>1</sup> The disease significantly impacts Australia's healthcare system, educational institutions, the workforce and the broader economy. CHD is one of the leading causes of death and hospitalisation of infants.<sup>2 3 4</sup> CHD is a chronic condition that, for many patients, requires complex, specialised care across the life course.<sup>5</sup>

CHD survival rates have improved dramatically due to medical advances. Most people with CHD now survive into adulthood, and currently adults living with CHD outnumber children with the disease.<sup>6 7 8 9</sup> The longer survivorship of people with CHD means we must now look beyond childhood and consider whole-of-life care and the cost to patients, the health system, workforce and national economy.

Despite the prevalence of CHD and its significant impact on individuals, families, the health system and society, there is a lack of awareness of CHD in the general community and among health professionals. There is also a lack of evidenced-based information and support for individuals with CHD.

Some sub-populations of Australians with CHD are disproportionately impacted by the disease, specifically: Aboriginal and Torres Strait Islander people; adolescents and young adults aged 15-24 years who are transitioning from paediatric to adult cardiac health services; and people living in remote, or rural and regional locations.

This Action Plan is the first nationally-coordinated effort to tackle CHD. It has been developed through close collaboration and consultation with clinicians, researchers, 'heart kids', parents and carers of 'heart kids', policymakers and members of the community. The Action Plan provides direction and guidance to transform care so that it is lifelong, holistic, person-centred and family-centred.



The Action Plan presents a comprehensive overview of the Australian CHD landscape, identifying needs, emerging challenges, opportunities for the CHD community and a Framework for Action.

Implementation of the Action Plan will deliver:

- the first ever Australian standards of care for CHD, including world first neurodevelopmental and mental health standards;
- more specialist CHD health professionals and specialist CHD centres to provide access to CHD-relevant health and allied health services and support for all Australians impacted by CHD irrespective of where they live;
- increased awareness of CHD and its unique challenges and improved access to information, support and resources for all people impacted by CHD;
- research evidence that will inform earlier interventions and improved therapies to reduce the burden of CHD on all those affected; and
- monitoring and surveillance of the implementation of the Action Plan to ensure benefit for those impacted by CHD and the wider community.

The Framework for Action (Appendix 1) shows the link between the three priorities, seven focus areas, 29 recommended actions and desired outcomes of the Action Plan.

The priorities and focus areas are:

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

## ACRONYMS

ACHD	Adult congenital heart disease
AIHW	Australian Institute of Health and Welfare
ARF	Acute rheumatic fever
CACH	Comprehensive Adult Congenital Heart Centres
CHAANZ	Congenital Heart Alliance of Australia and New Zealand
CHD	Childhood heart disease which includes congenital and acquired heart disease
COCOON	Circle of care optimising outcomes for newborns
CQR	Clinical quality registries
CSANZ	Cardiac Society of Australia New Zealand
ESSA	Exercise and Sports Science Australia
FTE	Full-time equivalent
GP	General practitioner
ICU	Intensive care unit
MDT	Multidisciplinary team
MRFF	Medical Research Future Fund
NATA	National Association of Testing Authorities
NDD	Neurodevelopmental disability
NDIS	National Disability Insurance Scheme
NHMRC	National Health and Medical Research Council
NP	Nurse practitioner
PATS	Patient Assistance (or Assisted) Travel Scheme
PICU	Paediatric intensive care unit
PROMs	Patient- and family-reported outcome measures
PTSD	Post-traumatic stress disorder
RACH	Regional Adult Congenital Heart Centres
RFS	Rheumatic Fever Strategy
RHD	Rheumatic heart disease
RHOF	Rural Health Outreach Fund
RN	Registered nurse
WHO	World Health Organization

A glossary of terms is provided in Appendix 2.

# CHILDHOOD HEART DISEASE (CHD) AND THE EMERGING CHALLENGES

## *BACKGROUND*

The 2011 CHD in Australia White Paper,<sup>10</sup> developed and released by HeartKids, reviewed the delivery of healthcare in Australia to children with CHD, both acquired and congenital. It found there were significant areas of deficit within service delivery and family support and outlined recommendations for improved healthcare delivery and support for CHD patients, their families and carers. Now, almost a decade later, many of these issues retain currency and are similarly under focus internationally. Action to redress under-resourcing of CHD care is critical.

This evidence-based Action Plan draws on domestic and international research and advice from experts in Australian national hubs of excellence. The Action Plan has been developed within a broad health policy environment and aligns with, complements and supports key national strategies and initiatives including the:

- policy directions in the National Strategic Framework for Chronic Conditions,<sup>11</sup> which provides the overarching national policy for the prevention and management of chronic conditions in Australia;
- recently-expanded Rheumatic Fever Strategy for the prevention and management of acute rheumatic fever (ARF) and rheumatic heart disease (RHD);
- renewed commitment to closing the gap for Aboriginal and Torres Strait Islander people;
- development of a national clinical quality registry strategy to improve clinical care and patient outcomes;
- development of Australia's future workforce; and
- Department of Health's Rural Health Outreach Fund (RHOF) to improve outcomes for people living in regional, rural and remote areas.

## *CHD IS A COMPLEX CHRONIC CONDITION*

CHD is a general term for a range of conditions that affect the normal workings of the heart and begin in childhood. There are many different heart conditions that can occur in childhood, with the main distinction being those present from birth (congenital) and those that develop during childhood (acquired).<sup>12</sup> Congenital forms of the disease are a leading cause of hospitalisations and death of Australian babies.<sup>13 14 15</sup>

CHD is a chronic condition that for many patients requires complex, specialised care across the life course.<sup>16</sup> It is fundamentally different from heart disease diagnosed in adult life, which typically occurs as arteries become blocked and heart valves deteriorate with age. Lifestyle factors often contribute to poor heart health in adulthood, but most CHD is unrelated to lifestyle.

## Congenital heart disease

Congenital heart disease is any defect of the heart structure or major blood vessels that is present at birth.<sup>17</sup> Some defects are mild and cause no significant disturbance to the way the heart functions;<sup>18</sup> however, more than half of all children with congenital heart disease will require treatment at some stage of life.<sup>19 20</sup> The causes of congenital heart disease remain largely unclear.<sup>21</sup> It is estimated that approximately 30 per cent of cases have a genetic cause or can be attributed to known genetic disorders; however, most cases are thought to arise through complex combinations of genetic factors, environmental factors and factors relating to maternal health.<sup>22</sup>

While there is no evidence for effective interventions to prevent congenital heart disease and no cure, treatment which can reduce the impact of the disease is available for most congenital heart defects. More than half of all children with a heart defect have a condition that is serious enough to require treatment.<sup>23</sup> Evidence is emerging that childhood surgical repair of the heart has longer-term impacts<sup>24 25 26 27 28</sup> and that early diagnosis and treatment can lead to better long-term management and health outcomes, including quality of life.<sup>29</sup> Surgery can reduce the impact of congenital heart disease, although it does not provide a complete cure, and patients have complex, life-long needs. Most cases of congenital heart disease have no known cause.<sup>30</sup>

## Acquired heart disease

Acquired heart disease refers to conditions developing in the heart which appears structurally and functionally normal at birth. These may include new conditions acquired after birth, such as:

- rheumatic heart disease (RHD), caused by an abnormal immune reaction to streptococcus A (strep A), a bacterial infection of the skin or throat. Strep A infections are more common in settings of household crowding, poverty, and where there is limited access to health hygiene infrastructure and medical services.<sup>31</sup> In Australia, 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.<sup>32</sup> Pacific Islanders and migrants from countries with a high-prevalence of strep A<sup>33 34</sup> are also at high risk,<sup>35</sup>
- viral myocarditis, impacting on heart muscle function;<sup>36 37 38</sup>
- Kawasaki Disease, where hyper-response to various infective triggers produces inflammatory changes across the body, including the coronary arteries;<sup>39 40 41</sup>
- genetic conditions where the heart appears normal initially but programmed genetic abnormalities produce changes in the heart, such as in cardiomyopathy and certain arrhythmias, as well as the early stages of adult heart disease;<sup>42</sup> and
- arrhythmias where the electrical control of the heart beat is abnormal. This mostly occurs for the first time after birth (occasionally before birth) and may have a variety of causes which include developmental abnormality of the electrical system, genetic abnormalities, and rhythm abnormalities occurring after intervention for congenital heart disease.<sup>43 44 45</sup>

Rheumatic heart disease, an acquired heart disease, is entirely preventable.<sup>46</sup>

## *THE HEALTH, SOCIAL AND ECONOMIC IMPACTS OF CHD*

CHD directly impacts over 65,000 Australians living with the condition, as well as their family members and carers.<sup>47</sup> CHD also impacts Australia's healthcare system, educational institutions, the workforce and the broader economy.<sup>48 49</sup> The economic burden of disease, including on education, employment and insurability, is now being recognised for adults living with CHD.<sup>50</sup>

Despite the prevalence of CHD and its significant impact on individuals, families, the health system and society, there is a lack of awareness of CHD in the general community and amongst health professionals, as well as a lack of evidence-based information and support for individuals with CHD.<sup>51</sup>

### **BOX 1: THE HEALTH, SOCIAL AND ECONOMIC IMPACTS OF CHD**

Over 65,000 Australian children and adults live with CHD, and their many family members and carers are also impacted.<sup>52</sup>

Congenital heart defects are the most common types of birth defects; in Australia, eight babies are born with a congenital heart condition every day.<sup>53</sup>

Congenital heart disease is one of the leading causes of death and hospitalisation in infants.<sup>54 55 56</sup>

Four young lives are lost every week.<sup>57</sup>

Aboriginal and Torres Strait Islander people experience higher rates of congenital and acquired heart disease and worse outcomes compared with non-Indigenous Australians.<sup>58 59 60</sup>

People with CHD and their families have 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).<sup>61 62 63 64 65 66</sup>

People with CHD are at a greater risk of neurodevelopmental impairment and disability including developmental delay and other learning difficulties.<sup>67</sup>

CHD poses a substantial economic burden on people affected by the disease, Australia's healthcare system, educational institutions, the workforce and the broader economy.<sup>68 69 70 71</sup>

There is a lack of awareness of CHD in the general community and among health professionals<sup>72</sup> and a lack of evidence-based information and support for individuals with CHD, their families and carers.<sup>73</sup>

## *THE CHANGING LANDSCAPE OF CHD AND THE EMERGING CHALLENGES*

Thanks to medical advances in Australia and internationally over the past 75 years, CHD survival rates have improved dramatically. Many childhood heart problems that were previously fatal are now chronic medical conditions requiring ongoing management. The decline in mortality has shifted over the past two decades and 95 per cent of people with CHD now survive into adulthood, but life expectancy continues to be below average.<sup>74 75 76 77</sup> Currently, adults living with CHD outnumber children with the disease, yet a lack of definitive research on the burden of disease suggests the number of people living with CHD could be far higher than current estimates.<sup>78</sup> This large and growing population requires lifelong and highly specialised medical care.



The gains in survival for CHD are a triumph, however, they bring new challenges. 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, education system, workforce, and national economy. There remains a focus on childhood, where the intensity of interventions occurs. Approximately half of all children with congenital heart disease and a significant lesion have surgery/catheter intervention within their first year of life.<sup>79</sup> This means that we must now look beyond childhood and consider whole-of-life care, from diagnosis through to infancy, childhood, adolescence, and adulthood.

The rapid growth in the number of adults living with CHD is placing 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; and
- workforce shortages.

These challenges are limiting patient access and compromising quality of care. It is critical that Australia's health system is shaped to recognise and value the needs of individuals, their families and their carers, and that person-centred and family-centred approaches underpin CHD care.

Further detailed information and references on the nature and prevalence of CHD, the health, social and economic burden of CHD and the emerging challenges of CHD are provided in Compendium C – Evidence.

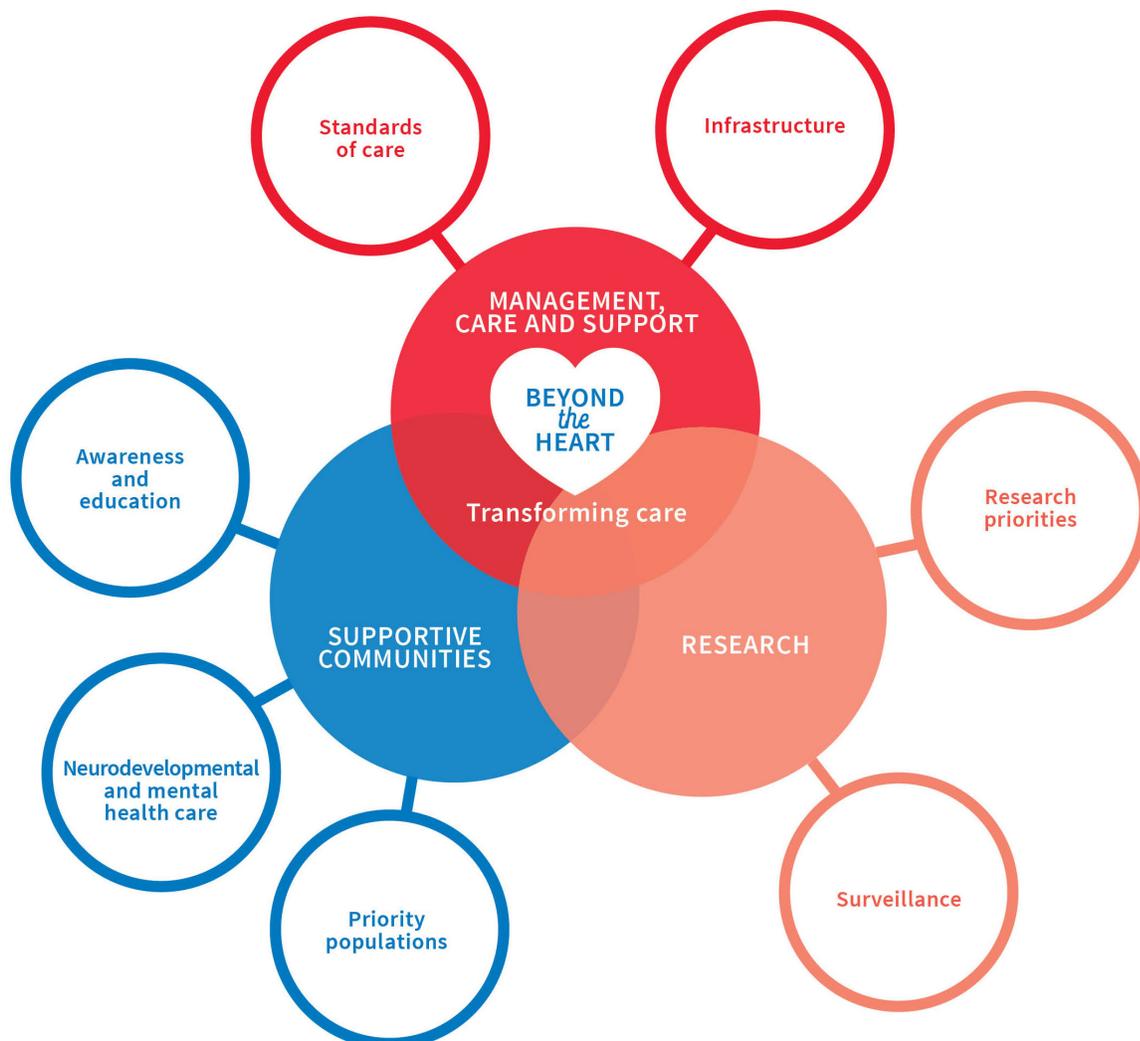
## THE GOAL AND THE APPROACH

The goal of the Action Plan is that people with CHD live longer, healthier and more productive lives through effective management of CHD across the life course.

This goal will be achieved by focusing on three priorities and seven focus areas:

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

*Figure 1: National Strategic Action Plan for CHD 2019 - Priorities and Associated Focus Areas*



## The importance of partnerships

Our approach to implementing the recommended actions in the Action Plan and achieving tangible outcomes for CHD patients and the CHD community in Australia will be based on the guiding principles and enablers of the National Strategic Framework for Chronic Conditions.<sup>80</sup> Implementation of the Action Plan will be informed by evidence-based practices and other related strategic health priorities. We will also develop and maintain key partnerships with those impacted by CHD, government organisations, the health and education sectors, advocacy and support groups, the research community and other industry organisations and agencies (see Compendium A – Consultation Summary and Implementation Partners for further information). People with CHD and their parents, family members and carers should be at the centre of all partnerships.

The effective prevention and management of childhood heart disease is strongly influenced by the contributions made by a wide range of Partners. These Partners include:

- individuals, carers and families;
- communities;
- all levels of government;
- non-government organisations;
- the public and private health sectors, including all health care providers and private health insurers;
- industry; and
- researchers and academics.

All Partners have shared responsibility for health outcomes according to their role and capacity within the health care system. Greater cooperation between Partners will lead to more successful individual and system outcomes. Actions included in this Action Plan are intended to guide Partner investment in the prevention and management of childhood heart disease and should be implemented collaboratively to achieve the best health outcomes.

## PRIORITY 1: MANAGEMENT, CARE AND SUPPORT

### *FOCUS AREA 1: STANDARDS OF CARE*

#### DESIRED OUTCOME

Australia will have its first national CHD standards of care including world first neurodevelopmental and mental healthcare standards.

#### WHY?

**Standards of care** are components of care that health professionals and health services should offer patients for a specific clinical condition. Standards define and uphold the safety and quality of healthcare provision and provide a framework to ensure people receive the best possible care and outcomes.<sup>81</sup> They guide delivery of care and the related service and resource requirements. Standards of care are best implemented on a national basis for areas of special need.

Australia has never had nationally agreed standards of care for CHD. Such standards are an important factor in the provision of seamless congenital cardiac care pathways. Without such standards, there are risks, including:

- inconsistency of practice and resourcing of patient care, particularly disadvantaging people with CHD in rural, remote and isolated communities;
- current care delivery processes can be fragmented and focused on 'moments of care' rather than lifelong 'continuity of care';
- lack of alignment between workforce and infrastructure planning and resourcing to meet the changing workforce needs (surgical, clinical care and allied healthcare) and infrastructure needs (facilities, technology and equipment) of CHD patients, their families and carers; and
- disconnect of patients transitioning from paediatric to adult CHD care, where over half of Australians with CHD are being 'lost to care',<sup>82 83</sup> and associated consequences for the appropriate management of their disease, mental health, well-being and life expectancy.<sup>84</sup>

While there are international and national best practice examples of congenital heart disease standards of care<sup>85 86</sup> and the Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease,<sup>87</sup> significant gaps exist. The development of the first Australian standards of care for CHD will be informed by relevant standards, statements and guidelines.

The development of the Australian standards of care for CHD will be overseen by a CHD taskforce comprising a working group of clinicians, researchers, consumers, and policy makers including representatives from Federal, State and Territory Governments. The standards of care will inform the development of:

- models of care;
- care pathways;
- clinical practice guidelines;
- workforce planning, including training of specialists, general practitioners (GP), nurses, allied health professionals and patient and family support workers; and

- infrastructure planning to assist in achieving alignment between practice and the standards of care.

#### WHAT WILL BE DIFFERENT?

- People with CHD will receive care of consistently high quality that is sustainable for the future.
- Improvements in clinical care will occur due to the provision of service and workforce standards that can be used when benchmarking and monitoring performance.
- Improved health outcomes will be achieved for people with CHD and their families, including quality of life, physical health, mental health and wellbeing, and neurodevelopmental outcomes.
- Lifelong opportunities for excellence in care will exist, resulting in a reduced burden of CHD on the healthcare system, educational institutions, workforce and the broader economy.
- A CHD taskforce will be established to oversee and guide the development of Australia’s first CHD standards of care.

MANAGEMENT, CARE AND SUPPORT – FOCUS AREA 1: STANDARDS OF CARE	
ACTIONS	DESCRIPTION
1.1 Develop national standards of care for CHD	<p>Develop a set of standards and service specifications that cover the entire patient pathway, from prevention, diagnosis, treatment, in-home care and end-of-life care, to make sure that every child, young person and adult with CHD, in every part of the country, will receive the same high standard of care.</p> <p>Standards of care should consider Australia’s unique geographical challenges and the needs of priority populations, and include:</p> <ul style="list-style-type: none"> <li>• organisation of care to meet current and future population needs;</li> <li>• service model;</li> <li>• care pathways to map the patient journey across primary, secondary and tertiary levels of care and community settings;</li> <li>• service specification standards and resource requirements, including personnel and equipment;</li> <li>• resources and frameworks for minimum effective communication with patients, families, and other care providers, including priority populations;</li> <li>• information reporting requirements including patient satisfaction measures and clinical outcomes measures; and</li> <li>• training, education and certification requirements for those involved in providing care, including clinicians, nurses, allied health practitioners, and patient and family support personnel.</li> </ul> <p>Establish a framework and processes for ongoing surveillance, review and periodic updates of the standards to ensure currency, relevance and links to related standards.</p>

<p>1.2 Develop authoritative national clinical practice guidelines on CHD for health services and health practitioners throughout Australia</p>	<p>Following the development of standards of care, a CHD taskforce will work with experts to devise a roadmap and priorities for the development of national clinical practice guidelines.</p> <p>The development of the national clinical practice guidelines for CHD will follow NHMRC guideline standards. The development of clinical practice guidelines (affirmed by CSANZ) will include a review of existing international clinical guidelines such as the American College of Cardiology/ American Heart Association’s Guidelines for the Management of Adults with Congenital Heart Disease and other international guidelines on CHD for the feasibility of their adaption and adoption and for consideration in the Australian context.</p> <p>To ensure currency and relevance, clinical practice guidelines will be developed with expert input and collaboration with the Australian Guideline Developers Network and other sources. Development of the guidelines will draw on the best evidence possible and input from public consultation, on conditions or procedures where evolving experience and research are altering strategies for care, require higher levels of uniform care, or where uncertainty exists regarding the choice of care modalities. Clinical practice guidelines will promote integrated care for all stages of the care pathway and deliver quality and consistency for current and future generations.</p> <p>The development of guidelines for echocardiographic screening for RHD is one priority area for consideration.</p> <p>Guidelines should be available in multiple formats, including web-based, to maximise accessibility and uptake by the multidisciplinary care team.</p>
<p>1.3 Establish a CHD taskforce to oversee the development of standards of care for CHD</p>	<p>Establish a CHD taskforce, comprising a working group of clinicians, researchers, consumers, and policy makers including representation from Federal, State and Territory Governments, to oversee and guide the development of:</p> <ul style="list-style-type: none"> <li>• the Australian standards of care for CHD, incorporating Mental Health and Neurodevelopmental Standards;</li> <li>• a roadmap and priorities for clinical practice guidelines aligned to the standards of care for CHD;</li> <li>• clinical practice guidelines in accordance with NHMRC guideline standards; and</li> <li>• workforce and infrastructure plans aligned to the standards of care.</li> </ul> <p>The CHD taskforce would also have a pivotal role in ensuring the monitoring and evaluation of the Action Plan’s progress and implementation.</p> <p>The CHD taskforce should be resourced with capacity to respond to areas of need identified during the development of the standards of care, and to address gaps between the care defined by the standards of care and current practice (e.g. development of models of care).</p>

## FOCUS AREA 2: INFRASTRUCTURE

### DESIRED OUTCOME

Australia will have a health system infrastructure which provides continuity of specialised care for CHD patients from birth through adolescence to adulthood.

### WHY?

**Infrastructure** refers to the service and resource requirements needed to deliver quality care for people with CHD who have complex health needs and require lifelong care from workforce operating from a range of institutions, both public and private.

Infrastructure includes the:

- the multidisciplinary workforce providing care; and
- the facilities, services and equipment that are needed for safety and excellence in CHD care.

Australia's current infrastructure is not consistent with international guidelines with impacts on service provision and health outcomes for Australians with CHD.<sup>88</sup> This includes a burden on the healthcare system, educational institutes, the workforce and broader economy.<sup>89</sup> CHD-relevant infrastructure in Australia remains under-resourced.<sup>90</sup>

The lack of adequate infrastructure can have a catastrophic impact at certain stages of the care journey, such as access to critical care facilities in childhood, or transition of care from paediatric to adult cardiac health services.<sup>91 92 93</sup>

### Workforce

A critical issue is the shortage of adult congenital heart disease (ACHD) cardiologists. In Australia, there are fewer ACHD cardiologists, surgeons and other specifically trained ACHD healthcare professionals than recommended by international standards.<sup>94</sup> In 2010, there were approximately 30 paediatric cardiologists in Australia,<sup>95</sup> yet the equivalent of only six full time cardiologists care for adults with CHD.<sup>96 97 98 99</sup>

Such lack of workforce infrastructure disproportionately impacts some population groups, such as Aboriginal and Torres Strait Islander people and those living in regional, rural and remote areas, and contributes to growing inequities in healthcare access and health outcomes. While investments have been made through the Rural Health Outreach Fund and Bonded Programs to better target medically trained professionals to regional, rural and remote areas, additional specialist-trained health professionals are required.<sup>100</sup>

There is a strong need for point-of-care staffing, and an increasing demand for certain professions across the life course, including psychologists, developmental specialists, genetic counsellors, nurses and transition care coordinators. This is particularly important for Aboriginal and Torres Strait Islander people and those living in regional, rural and remote areas. No dedicated ACHD Centres in Australia have an attached psychologist or physiotherapist, and most have no social worker or even a nurse educator.<sup>101</sup>



Additional workforce related issues include the opportunity for training pathways and positions for health professionals, as well as a low level of CHD awareness, knowledge and capacity amongst primary care providers.

The new standards of care for CHD will inform workforce planning and development. Focus Area 5: Priority Populations, discussed later in this Action Plan, contains further recommendations for infrastructure investments that will have an immediate and significant impact on Aboriginal and Torres Strait Islander communities and other priority populations.

### *Facilities, services and equipment*

CHD services in Australia are severely under-resourced, particularly in terms of dedicated facilities and family support for both paediatric and adult services, although it is particularly pressing for adult services due to the large and growing cohort of adults with CHD.<sup>102</sup>

Consistent with international best practice, a hub-and-spoke model of care, comprising centralised comprehensive adult congenital heart (CACH) centres to provide care for people with moderate to severe spectrum of disease, and regional adult congenital heart (RACH) centres for the milder spectrum of disease, is recommended for CHD care.<sup>103 104 105 106</sup> There are currently only five designated CACH centres across Australia and no RACH centres. Based on international best practice of one centre for every two million population,<sup>107</sup> Australia should have at least 12.5 CACH centres based on a population of 25 million.

The inadequate number of adults centres as well as insufficient resources in paediatric intensive care units (PICU) and intensive care units (ICU) places pressures on the available dedicated centres and burdens patients and families.<sup>108</sup> Patients and families incur major travel and accommodation costs, insufficient family facilities in hospitals, lack of access to respite care, and surgery cancellations. Greater co-location of family support services within hospital settings would help to alleviate some of these issues. Additionally, the adaptation and use of e-health technologies (i.e. video conference) for treatment and provision of information and support would assist in reducing the needs of rural and remote patients to travel to specialist centres.

Improved forward planning to address the emergence of new treatment options, i.e. personalised medicine, and the associated infrastructure costs over the next decade can have great impact on patient survival and quality of life.<sup>109</sup>

### **WHAT WILL BE DIFFERENT?**

- The health workforce will be trained, resourced and distributed to meet identified needs.
- The health workforce will be working to its full scope of practice.
- Effective multidisciplinary teams and healthcare partnerships will be in place.
- Service delivery will be coordinated and complete.
- There will be improved health outcomes including quality of life, physical and mental health, and neurodevelopmental outcomes for all people with congenital heart disease and their families.
- The disparity in health outcomes due to CHD between Aboriginal and Torres Strait Islander people and non-Indigenous Australians will be reduced.
- The disparity in health outcomes due to CHD between people living in regional, rural and remote areas and metropolitan areas will be reduced.

## MANAGEMENT, CARE AND SUPPORT - FOCUS AREA 2: INFRASTRUCTURE

ACTIONS	DESCRIPTION
2.1 Develop a CHD infrastructure plan	<p>Develop a national CHD infrastructure plan to ensure Australia can provide quality and accessible care to meet current and future population needs.</p> <p>The infrastructure plan will:</p> <ul style="list-style-type: none"> <li>• define a multidisciplinary care team for both paediatric and adult care settings;</li> <li>• identify the current workforce composition and services, dedicated facilities and family support across Australia and compare this to the specifications identified in the standards of care to identify resource gaps and develop a workforce plan to highlight investment priorities; and</li> <li>• review existing training (vocational, undergraduate and postgraduate, and fellowship) to identify how future education, training and certification can improve health outcomes and meet the evolving needs of people with CHD. Opportunities include funded fellowships, training in congenital cardiac surgery, and education, training and certification requirements for those involved in providing care.</li> </ul> <p>The national standards of care for CHD (Focus Area 1 of this Action Plan) will form the basis of infrastructure planning and provide guidance for investment and action by Australian, State and Territory Governments.</p>
2.2 Invest in technological infrastructure	<p>Invest in innovative strategies to use technology to broaden access to health services, including appropriate use of telehealth and digital health options, and facilitate sharing of health information between the workforce and patient, including use of the My Health Record.</p> <p>Develop integrated digital platforms for effective and efficient capture of patient and family reported outcomes (PROMs) in care services to assist informed and shared clinical decision-making and care.</p>

<p>2.3 Deliver education, training and support for health professionals</p>	<p>Develop the workforce and its capacity to provide consistent, quality care for people with CHD through education, training and support focused on:</p> <ul style="list-style-type: none"> <li>• patient and family-centered care for people with CHD;</li> <li>• the physical, psychological, social and cognitive impacts of CHD;</li> <li>• co-morbidities, including neurological impairment;</li> <li>• culturally safe and appropriate communication and counselling techniques;</li> <li>• support for health professionals to minimise burnout and compassion fatigue;</li> <li>• referral pathways;</li> <li>• improving knowledge of primary care doctors, nursing staff and general cardiologists about the special needs of this group; and</li> <li>• improving knowledge of nursing, allied health practitioners as personnel providing patient services and support about the needs of CHD patients, families and carers.</li> </ul> <p>Deliver education programs to support primary care staff, rural health professionals and community-based allied health practitioners and address the needs of workers that support priority populations (Focus Area 5 of this Action Plan) including the development of culturally appropriate and accessible education for Aboriginal and Torres Strait Islander health practitioners.</p> <p>Develop resources and tools to support health professionals including nursing staff to deliver whole-of-life care and secure best patient outcomes (e.g. map CHD specialist services for regional clinics to support timely and appropriate referral).</p> <p>Engage the broader cardiology community and cardiologists working in both the public and private system in the development of education, training and support.</p> <p>Align education, training and support with standards of care for CHD.</p>
<p>2.4 Support the management of CHD in primary care</p>	<p>Develop resources, including reviewing existing chronic disease management mechanisms such as general practitioner management plans, nursing care provision, team care arrangements and mental health plans, to ensure:</p> <ul style="list-style-type: none"> <li>• support for general practitioners, registered nurses and Aboriginal and Torres Strait Islander health workers in the management of CHD patients of all ages and their families and carers; and</li> <li>• access to appropriate support and holistic care for CHD patients of all ages and their families and carers.</li> </ul>
<p>2.5 Disseminate best practice information and approaches throughout services in Australia</p>	<p>Expand and support the CHAANZ network and other communities of practice around CHD for health professionals, to share information and support each other in their practice.</p>

## PRIORITY 2: SUPPORTIVE COMMUNITIES

### *FOCUS AREA 3: AWARENESS AND EDUCATION*

#### DESIRED OUTCOME

People with CHD and their families will have access to information, support and resources when and where they are needed.

#### WHY?

**Awareness and Education** includes the range of evidence-based communications, information and learning approaches often used in health promotion, health education and health literacy to empower all members of the CHD community.

This Action Plan recognises the importance of timely and readily-available access to evidence-based, culturally sensitive, quality information and support. It also highlights the importance of the broader community being aware of the condition and its implications.

Although information and support are currently available, these may not be evidence-based, appropriately targeted or resourced. Parental information on CHD is insufficient,<sup>110</sup> particularly for priority populations with unique cultural needs.<sup>111 112</sup> In addition, information sources such as the valued but under-resourced HeartKids 1800 Line, are not consistently accessible nationally.

There is a need for a more comprehensive, coordinated and better-resourced approach to awareness and education targeted at:

- people impacted by CHD including individual patients and their families and carers; and
- the wider community, for example, education and employment settings, sporting organisations, and common services such as housing, insurance and finance.

#### *People impacted by CHD – individual patients and their families and carers*

People impacted by CHD need to access information and support in a timely manner, regardless of where they are located. This is critical not only when a child is first diagnosed with CHD but throughout the patients' life.<sup>113 114 115</sup>

Lack of support is an identified issue for bereaved families. Beyond peer support and connection with others in similar situations, there is no specific service or counselling available to families who have lost a child to CHD.

Peer support programs, such as those provided by HeartKids, have been shown to significantly improve the quality of life of all people impacted by CHD.<sup>116 117</sup>

#### *The wider community*

Research has highlighted the need for the education of patients, families, carers and the community more broadly. Community settings, such as schools, workplaces, financial institutions and sports and recreation groups offer the opportunity to provide information about support and required intervention



to assist people to navigate the system, as well as to provide equity of access and prevent discrimination.<sup>118 119</sup>

### WHAT WILL BE DIFFERENT?

- People impacted directly by CHD will have increased access to information and support that prepares them to manage their condition, informs choices and provides clear pathways of care for their individual healthcare needs, across the lifespan.
- Families and carers will have improved access to quality, evidence-based materials that equip them to understand their child's condition, know what to expect and how, when and where to access support to achieve the best outcomes, for their child and themselves.
- People with CHD will have improved opportunities to grow and develop in a supportive community that understands the impacts of the condition and delivers appropriate and relevant solutions to maximise and support quality of life.

## SUPPORTIVE COMMUNITIES – FOCUS AREA 3: AWARENESS AND EDUCATION

ACTIONS	DESCRIPTION
<p>3.1 Increase access to quality, relevant information, education and support through a range of approaches, phone, face to face and digital.</p>	<p>Fund services that enable timely access to information and services for people impacted by CHD, including their families, carers, health professionals and the broader community.</p> <p>Build on existing 1800 HeartKids telephone service to ensure capacity to meet community demand in a timely manner, increasing reach and access across the country. This type of service would provide:</p> <ul style="list-style-type: none"> <li>• immediate support and referral to local support team member;</li> <li>• provision of information in relation to a query; and</li> <li>• connection/referral to services and programs (internal and external).</li> </ul> <p>Building on existing resources, invest and implement a digital strategy that provides a web-based CHD education portal, linkages to key parent sites, developed in consultation with CHD consumers. This would include:</p> <ul style="list-style-type: none"> <li>• development of a range of evidence-based, quality information and education materials to improve accessibility of information in response to a range of health literacy needs and learning styles, for example, development of video stories, and a range of educational webinars with experts;</li> <li>• linkage to parent education, parent resource sites; and</li> <li>• building opportunities to link CHD consumers to allied health services.</li> </ul> <p>Consider development of a national publicly-accessible register of qualified professionals and local and national peer groups to improve access to clinicians and other health professionals with specific CHD skills.</p> <p>Invest in the development, through consultation, of culturally appropriate communications, resources and support materials that are accessible and relevant to all people impacted by CHD, with a particular focus on Aboriginal and Torres Strait Islander people.</p> <p>In consultation with existing stakeholders, such as RHD Australia and peak bodies in Indigenous health, map existing resources, identify gaps and needs and determine how these needs can be met and a response developed.</p>
<p>3.2 Increase access to ongoing support by funding inpatient and outpatient community (peer) support team members</p>	<p>Establish community/peer support teams based in hospital and community settings to provide support to individuals and families working alongside the clinical team to:</p> <ul style="list-style-type: none"> <li>• provide face-to-face support to families ensuring connected care before and after hospital admissions;</li> <li>• facilitate connection to HeartKids community, peers as mentors, parents as advocates;</li> <li>• provision of HeartKids programs and services tailored to family/community need; and</li> <li>• liaison between CHD cardiac coordinators (see Focus Area 5: Priority Populations in this Action Plan), health service providers and cardiac social workers.</li> </ul>

## SUPPORTIVE COMMUNITIES – FOCUS AREA 3: AWARENESS AND EDUCATION

ACTIONS	DESCRIPTION
<p>3.3 Increase the number of young people engaged with transition and ongoing specialist CHD care</p>	<p>Fund specific digital strategies to increase the number of young people engaged with transition and ongoing specialist CHD care.</p> <p>In conjunction with young people through HeartKids youth advisory councils, develop a digital support strategy, for example, an evidence-based app in collaboration with a multidisciplinary research team that assists in understanding condition, self-management, attending appointments, engaging with allied health and psychological care during transition and beyond. This would align with clinical transition processes.</p> <p>Fund ongoing projects that facilitate connection with peers, support personal growth, self-management and leadership opportunities.</p> <p>Secure funding for camps aimed at young people to ensure access to this valuable opportunity, and ensure this includes priority populations, such as young Aboriginal and Torres Strait Islander people, with appropriate resourcing for Indigenous health and social workers.</p>
<p>3.4 Deliver awareness and education for non-health professionals</p>	<p>Fund a community-based awareness campaign to improve understanding of CHD and its impact across the whole lifespan. The campaign will focus on recognition of condition, impact on families, and seeking treatment and the need for lifelong care.</p> <p>Develop resources and policy guidance for key non-health institutions (e.g. schools and education settings) to create supportive community environments for people with CHD.</p> <p>Deliver awareness and education and provide support to professionals working outside the healthcare system, including community environments where people live, learn, work and play, e.g. Aboriginal community-controlled organisations, early childhood educators and teachers, sports coaches and home and community care workers.</p>

## FOCUS AREA 4: NEURODEVELOPMENTAL AND MENTAL HEALTHCARE

### DESIRED OUTCOME

People with CHD and their families will experience improved mental health, cognitive outcomes and quality of life as a result of world first neurodevelopmental and mental health standards.

### WHY?

**Mental healthcare for CHD** is defined as integrated, specialised and evidence-based care that focuses on reducing mental health burden and optimising emotional and psychobiological wellbeing for affected foetuses, infants, children, young people, adults and their families. There is an emphasis on prevention and early intervention, through evidence-based screening, assessment and psychological care and treatment, as part of routine cardiac care.

**Neurodevelopmental care for CHD** is defined as integrated, specialised and evidence-based care to prevent or minimise neurodevelopmental or neurocognitive impairment in infants, children, young people and adults with CHD, with an emphasis on early intervention, ongoing surveillance and specialised intervention, as indicated.

Worldwide attention is now being directed toward understanding the long-term quality of life and co-morbidities which impact it, including mental, cognitive and emotional health outcomes and their trajectory in this growing population.

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.<sup>120 121 122 123 124 125</sup> 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.

Significantly, the families of people with CHD, including parents, carers and siblings, also experience significant hardship and suffering.<sup>126 127</sup> People with CHD as well as their families have 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).<sup>128 129 130 131 132 133</sup>

Optimal care for CHD involves cardiac care that integrates neurodevelopmental and mental healthcare as core business.<sup>134 135</sup> An emphasis on earlier interventions and quality of life across the life course, beginning before birth, through infancy to adulthood, is required.<sup>136</sup>

## WHAT WILL BE DIFFERENT?

- Australia’s national standards of mental healthcare and neurodevelopmental care for CHD will ensure that neurodevelopmental and mental healthcare is a core part of CHD care and services, and subsequently, people with CHD and their families will experience improved mental health and cognitive outcomes and quality of life.
- People with CHD will have access to best practice neurodevelopmental and mental healthcare with a strong focus on optimising quality of life.
- The health and wellbeing of people with CHD and their families will be supported by a holistic approach to care that places equal emphasis on both physical and mental health.
- People with CHD and their families will be more empowered to seek help and self-manage as appropriate.
- The gap in quality of life, including neurodevelopmental, psychosocial and behavioural outcomes, between people with CHD and their peers, will narrow.

### SUPPORTIVE COMMUNITIES – FOCUS AREA 4: NEURODEVELOPMENTAL AND MENTAL HEALTHCARE

ACTION	DESCRIPTION
4.1 Develop standards of mental healthcare for CHD	<p>As part of the development of standards of care for CHD (Focus Area 1 in this Action Plan), develop world first national standards of mental healthcare for CHD. The standards of mental healthcare for CHD will:</p> <ul style="list-style-type: none"> <li>• build on existing good practice, including The Heart Centre for Children’s four-tier model of integrated mental health care;</li> <li>• cover the lifespan and the different issues faced at different life stages, including the mental health of expectant parents following fetal cardiac diagnosis, young people navigating the transitions of adolescence, children and adults awaiting heart transplantation, living with CHD into adulthood, and bereavement care; and</li> <li>• be person-centred and family-centred, and inclusive of parents, carers and siblings.</li> </ul> <p>Explore potential for transferability of standards of mental healthcare to other chronic conditions, such as cancer, asthma, neuromuscular diseases, other congenital anomalies, and other critical or chronic illnesses with childhood onset.</p>
4.2 Develop standards of neurodevelopmental care for CHD	<p>As part of the development of standards of care for CHD (Focus Area 1 in this Action Plan), develop national standards for neurodevelopmental care for CHD.</p> <p>The standards of neurodevelopmental care for CHD will:</p> <ul style="list-style-type: none"> <li>• build on existing good practice, including the Queensland Paediatric Cardiac Service,<sup>137</sup> and draw on international and Australian resources including the American Heart Association’s best practice recommendations<sup>138</sup>;</li> <li>• focus on early intervention and ongoing surveillance across the lifespan; and</li> <li>• address surveillance, screening, evaluation, diagnosis, and management of developmental disorders or disabilities in the paediatric congenital heart disease population.</li> </ul>

**SUPPORTIVE COMMUNITIES – FOCUS AREA 4: NEURODEVELOPMENTAL AND MENTAL HEALTHCARE**

ACTION	DESCRIPTION
4.3 Establish clear models of care	<p>Build on Queensland Paediatric Cardiac Service’s existing good practice model to develop and pilot a national long-term developmental care pathway. Pathways for developmental surveillance should reflect the following principles:</p> <ul style="list-style-type: none"> <li>• known risk: specifically, children undergoing cardiac surgery under 12 months of age, post transplantation, and survivors of extracorporeal life support (ECLS) are at greatest risk;</li> <li>• family-centred: services and support should be provided close to home; and</li> <li>• long-term: surveillance, screening and assessment into adolescence at recommended age points are required as deficits may emerge over time.</li> </ul> <p>Test developmentally-supportive practices in intensive care that incorporate developmental care and infant and parent mental health. Draw on good practice models such as the Royal Children’s Hospital’s Circle of Care Optimising Outcomes for Newborns (COCOON) model. The models of care will inform the development of the standards for neurodevelopmental care for CHD.</p>

## FOCUS AREA 5: PRIORITY POPULATIONS

### DESIRED OUTCOME

Australia will have a health system that is responsive to the specific needs of identified priority populations with CHD through provision of infrastructure and continuity of specialised care from birth through adolescence to adulthood.

### WHY?

**Priority Populations** are groups that are identified as a priority for action to prevent and manage chronic conditions. Some priority populations are disproportionately affected due to a complex interaction between the physical environment, social and cultural determinants and biomedical and behavioural risk factors.<sup>139 140</sup> Populations may be prioritised due to unique challenges associated with their chronic condition, barriers to accessing care, or an increased risk due to the presence of risk factors.

Priority populations in this Action Plan include Aboriginal and Torres Strait Islander people, young adults aged 15-24 years who are transitioning from paediatric to adult cardiac health services and people living in remote, or rural and regional locations. Barriers to effective treatment among these priority populations include:

- geographical remoteness,<sup>141 142 143</sup>
- lack of culturally appropriate healthcare services,<sup>144 145</sup>
- legal and institutional barriers to accessing support; and
- 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.

### WHAT WILL BE DIFFERENT?

- The health system at all levels will be responsive to the specific needs of priority populations to effectively address CHD by providing culturally safe and appropriate services, accessible health services that are effective, high-quality and affordable and flexible service options.
- Individualised and holistic transition support, care and coordination will be available to all adolescents and young adults to support their transition from paediatric to adult health services and reduce 'loss to follow-up' rates.
- Aboriginal and Torres Strait Islander people will partner in, and lead, the planning, design, evaluation and implementation of locally responsive and culturally appropriate services.
- The disparity in health outcomes due to CHD between Aboriginal and Torres Strait Islander people and non-Indigenous Australians will be reduced.
- Technology will broaden access to health services, including appropriate use of telehealth and digital health options.
- The capability of the health workforce will be enhanced to meet current and future needs of Aboriginal and Torres Strait Islander people and other priority population groups.
- Community empowerment and targeted action will improve local and population health outcomes for priority populations with CHD.

## SUPPORTIVE COMMUNITIES – FOCUS AREA 5: PRIORITY POPULATIONS

ACTIONS	DESCRIPTION
<p>5.1 Fund CHD cardiac coordinators in each jurisdiction</p>	<p>Fund CHD cardiac coordinators in each jurisdiction to coordinate and case-manage regional, rural and remote patients and families. This will include a focus on Aboriginal and Torres Strait Islander people and adolescent/young adult patients. The coordinator role will be at the centre of a multidisciplinary team-based model of care, incorporating:</p> <ul style="list-style-type: none"> <li>• registered nurse (RN) role, with the potential to train and convert to nurse practitioner (NP) where appropriate. A total 6.5 FTE is a minimum requirement, based on locations best-suited to serving priority populations, in particular, Aboriginal and Torres Strait Islander people, and allocated as follows: <ul style="list-style-type: none"> <li>○ Northern Territory 2.0 FTE based in Darwin and Alice Springs;</li> <li>○ Western Australia 1.5 FTE based in Perth and Kimberley;</li> <li>○ Queensland 1.5 FTE based in Cairns and Brisbane; and</li> <li>○ South Australia, New South Wales/Australian Capital Territory, Victoria/Tasmania 0.5 FTE each; and</li> </ul> </li> <li>• CHD cardiac coordinators to focus on: <ul style="list-style-type: none"> <li>○ provision of specialist outreach clinics;</li> <li>○ coordination of surgery and interventional procedures;</li> <li>○ development of care pathways, including transition pathway;</li> <li>○ reducing failure to attend rates and improving transition to adult cardiac care; and</li> <li>○ providing education to young people with CHD and their families and the health workforce.</li> </ul> </li> </ul>
<p>5.2 Fund cardiac sonographers and a portable echo machine in each jurisdiction</p>	<p>Rural/remote access to echocardiography (echo) is a priority. Echo is essential to CHD services and to the RHD program. Access to echo for young people living with RHD is needed for monitoring disease progression, assessing the duration of preventative treatment and making decisions about surgical timing. Echo is required for diagnosis, timing of surgery, pre and post-operative assessment, and long-term follow up of CHD and RHD.<sup>146</sup> There is also increasing use of echo for RHD screening in school-age children. Access to echo is limited in some regional and remote areas. Requirements include:</p> <ul style="list-style-type: none"> <li>• A total of 4.5 FTE cardiac sonographers allocated as follows: <ul style="list-style-type: none"> <li>○ Western Australia, Northern Territory and Queensland 1.0 FTE each; and</li> <li>○ South Australia, New South Wales/Australian Capital Territory, Victoria/Tasmania 0.5 FTE each; and</li> </ul> </li> <li>• Eight portable echo machines.</li> </ul>
<p>5.3 Evaluate current specialist outreach services for regional and remote communities, including remote Aboriginal communities, and fund outreach services where significant gaps exist</p>	<p>Evaluate current specialist outreach services for regional and remote communities, including remote Aboriginal communities and fund identified gaps in outreach services to reduce barriers to accessing services due to geographical disparity in services.</p> <p>Consider innovative and flexible service provision options such as e-health technologies, as a means of delivering information and services (e.g. telehealth), particularly for those in rural and remote areas where access is problematic. Use and expand telemedicine where appropriate.</p>

## SUPPORTIVE COMMUNITIES – FOCUS AREA 5: PRIORITY POPULATIONS

ACTIONS	DESCRIPTION
<p>5.4 Establish models of care for the transition of care from paediatric to adult cardiac health services, including investment in transition nurses</p>	<p>Draw on existing models of best practice, including the Royal Children’s Hospital in Melbourne, to develop and pilot a transition model of care that aims to reduce loss to follow-up rates and re-engage adults lost to care by addressing common reasons for ‘loss to follow-up’ including insufficient patient/carer preparation, inadequate service integration and limited access to adult CHD specialists.<sup>147 148</sup></p> <p>Fund transition nurses to support transition across paediatric and adult cardiac health services. Based on the number of people with CHD aged 15 to 25 years in each jurisdiction, this would equate to 12 FTE positions, based in paediatric and adult centres, as follows:</p> <ul style="list-style-type: none"> <li>• 4.0 FTE in Victoria;</li> <li>• 3.0 FTE in New South Wales;</li> <li>• 2.0 FTE in Queensland; and</li> <li>• 1.0 FTE each in South Australia, Western Australia and the Northern Territory.</li> </ul> <p>Partner with primary care and emergency departments to identify young adults who are lost to follow-up and reconnect them with specialist CHD care.</p>
<p>5.5 Investigate options for reducing barriers to accessing care and supporting people with CHD and their families</p>	<p>Provide support for people with CHD and their families to access and navigate existing services and government support including Carers Allowance, NDIS and Centrelink.</p> <p>Review disability and carer payments to ensure children and adults with CHD can access required support. Children and adults with CHD experience significant challenges in meeting the current NDIS access criteria despite the significant disability associated with the chronic conditions associated with CHD which substantially impact the everyday lives of those affected by CHD.</p> <p>Introduce a national patient travel and accommodation assistance scheme (PATS) to overcome jurisdictional differences in eligibility and operation. Increase PATS funding to alleviate the significant out-of-pocket costs of patients and their families and facilitate timely treatment and care.</p>

## PRIORITY 3: RESEARCH

### *FOCUS AREA 6: RESEARCH PRIORITIES*

#### DESIRED OUTCOME

Australia will lead the way in research to:

- Identify the factors that cause CHD, both genetic and environmental, to work towards reducing prevalence in the future;
- Identify the burden of neurodevelopmental, psychosocial, poor mental health and economic factors on patients with CHD in order to improve the quality of life of CHD patients and their families;
- Identify the health factors that impact CHD pre- and post-natally to inform evidence base and transform care;
- Understand Australian-relevant epidemiology to determine effective interventions; and
- Inform the development of an evidence-supported exercise prescription for patients with CHD.

#### WHY?

**CHD Research** is research into the causes, treatment and management of CHD and involves medical and health research.<sup>149</sup>

**Research Impact** is the contribution that research makes to the economy, society, environment or culture, beyond the contribution to academic research.<sup>150</sup>

**Consumer Engagement** in research means consumers, community members, researchers and research organisations working in partnerships, to improve the health and well-being of all Australians through health and medical research.<sup>151</sup>

The Research Expert Working Group of this Action Plan identified key research priorities that would have the greatest impact on the whole-of-life health and well-being of those impacted by CHD. Research priorities target key stages in the lives of patients, providing earlier diagnosis and optimised treatments, and maximising the benefits of successfully transitioning CHD children into the adult health system.

Research and surveillance are essential for:

- strengthening evidence-based practice for the diagnosis and management of CHD;
- identifying ways of preventing or curing CHD;
- understanding the burden and impact of the disease;
- informing health policy; and
- evaluating the effectiveness of efforts to reduce the physical, psychological and economic impacts of congenital heart disease; including the uptake and impact of this Action Plan.

Implementation of the research priorities will build and strengthen world-leading collaborations developed through the Congenital Heart Alliance of Australia and New Zealand (CHAANZ) and the



Australia and New Zealand Fontan Registry and draw on the expertise of many specialist health and research professionals that serve on the HeartKids Research Advisory Committee and HeartKids Health and Clinical Advisory Committee.

Working closely with HeartKids, these researchers and clinicians are uniquely positioned to gain years of improved quality of life for patients by innovating and optimising diagnosis, management and quality of care and ensuring meaningful consumer engagement and significant research impact.

#### WHAT WILL BE DIFFERENT?

- Evidence will inform clinical standards of care, and health and hospital system planning including workforce planning and optimal patient outcomes.
- New opportunities in neurological and mental health translational research will inform and reshape health and hospital systems, improve patient and family counselling and provide models of care transferable to other disease and chronic illnesses.
- Development and access to new interventions and treatment options will be accelerated.
- New strategies to remove barriers to adolescent and young adult clinical care will be identified, helping close the gap on this Action Plan priority.
- Sharing best practice across Australia and internationally will increase engagement and improve outcomes for this vulnerable patient cohort.
- There will be purposeful collaboration with researchers across health services and institutions, and with the private sector.
- There will be an identified model of monitoring and surveillance performance going forward.
- Consumers will be engaged in setting research priorities and measuring research impact for the benefit of consumers and the CHD community.

## RESEARCH – FOCUS AREA 6: RESEARCH PRIORITIES

ACTION	DETAIL
<p>6.1 Understand the genetic causes of, and predisposition to, CHD to provide support for those impacted by CHD</p>	<p>Advances in molecular genetics and genomics have increased our understanding of how we can use genetic information from next-generation sequencing technology (including whole genome sequencing) and clinical observations to determine the role of genetics in causing structural heart disease in neonates and children, and to provide information regarding recurrence risks in further pregnancies and offspring of affected individuals.</p> <p>Environmental and epigenetic factors are also implicated in causing CHD, and the ability to measure the impact of a range of factors will help determine the role of such environmental factors as a prelude to understanding the causes of altered cardiac development and establishing preventative strategies. A diagnosis can be important in the planning of future pregnancies, in clinical management of the patient and family, and in determining risk for offspring of the patient.</p> <p>Such information will, in the future, inform the development of clinical management and counselling strategies that are personalised according to the individual’s genotype. This would likely deliver benefits in perioperative management, drug dosing, prognostication and non-directive genetic counselling. Validation of genetic and environmental associations requires the assessment of a large number of patients who are followed prospectively and enhancement of the capacity of our health system to integrate precision approaches.</p> <p>Key deliverables:</p> <ol style="list-style-type: none"> <li>1. Assess the clinical and psychological value of providing National Association of Testing Authorities (NATA)-accredited sequencing and advanced genetic advice to families with multiple affected members and other relevant sub-groups, but who currently do not have access to clinical genetics services.</li> <li>2. Develop a strategy to provide whole genome sequencing information to clinicians as soon as practical after the diagnosis of CHD, often antenatally, and assess its value in complementing existing management pathways, defining risk and individualising care. This information would be available for early cardiac management, focused neurodevelopmental care and pharmacogenomic application.</li> </ol>

## RESEARCH – FOCUS AREA 6: RESEARCH PRIORITIES

ACTION	DETAIL
6.2 Prevent neurodevelopmental and mental health complications	<p>Many children with CHD experience neurodevelopmental and mental health complications. Particularly, those children who undergo major cardiac surgery as part of their treatment of CHD are at a major risk (up to 20 per cent) of developing a neurodevelopmental disability (NDD) including developmental delay, motor and cognitive impairment and later school-based issues.</p> <p>NDD has implications for future psychosocial adjustment, mental health and quality of life, as well as dependence on support from the healthcare system. Early identification of those children at risk of such complications can be achieved by prospectively assessing and screening neurodevelopmental abilities from birth. This can be done by using innovative genetic approaches to screen for genetic variants that may predispose children to NDD, or by identifying potential social and clinical observations correlated with risk, which may identify those children who would benefit from relevant interventions (including peer and community support programs, and psychological support) to improve resilience and reduce developmental impact and future reliance on the welfare system. In addition, the mental health of parents and family members in the prenatal period as well as after birth and early childhood has been shown to impact childhood neurocognitive, behavioural and emotional outcomes.</p> <p>Key deliverables:</p> <ol style="list-style-type: none"> <li>1. Collect and analyse data on neurodevelopmental impacts on children, and the mental health of patients and family members.</li> <li>2. Determine appropriate approaches, through genetic screening and identified clinical and social observations, for identifying those children at risk.</li> <li>3. Inform clinical trials to measure the benefits of identifying children with the greatest future risk of requiring community/welfare dependence, to test early interventions with children and families to improve resilience and lessen future reliance.</li> <li>4. Develop a research framework to establish the cost-effectiveness of such early interventions.</li> </ol>

## RESEARCH – FOCUS AREA 6: RESEARCH PRIORITIES

ACTION	DETAIL
<p>6.3 Understand the impact of prenatal and postnatal factors affecting the health of children born with CHD and their families</p>	<p>Most serious CHD is identified during pregnancy or within the first three months of life. We can identify individual characteristics both before and after birth that influence long-term outcomes for both child and family. However, we have not yet examined these in an integrated way between traditional silos such as obstetrics, neonatology, genetics, adolescent medicine and health economics.</p> <p>Prospective acquisition of clinical, social and genetic information from a cohort of children born with CHD will provide the opportunity to link characteristics with outcomes. This would provide access to numerous investigators from apparently-disparate fields and would significantly facilitate a number of research priorities and inform development of best practice. Such a cohort, followed indefinitely, will be of international significance.</p> <p>Key deliverables:</p> <ol style="list-style-type: none"> <li>1. Establish the framework and recruitment to a national cohort of neonates with CHD, diagnosed both pre- and postnatally and followed indefinitely.</li> <li>2. Make data from the prospective cohort widely available to researchers working in related and unrelated fields, to build a picture of the experience and impact of CHD on all Australian newborns and their families (medical, social, psychological, economic), including access to treatments and outcome data to inform best practice nationwide.</li> </ol>

## RESEARCH – FOCUS AREA 6: RESEARCH PRIORITIES

ACTION	DETAIL
<p>6.4 Measure longitudinal impact and inform practice with the National Congenital Heart Disease Registry</p>	<p>The key to improving the lives of those with CHD is to understand and track the variability in patients' conditions, treatments and outcomes over the life of each patient, to identify those interventions which have the most benefit, and to close the circle of transition of care from paediatric to adult healthcare.</p> <p>While there are some regional tracking systems, or registries, for infants and children, there are no national or international systems which track older children and adults with CHD. HeartKids has supported the establishment of the Congenital Heart Alliance of Australia and New Zealand (CHAANZ) to develop a pilot registry, building on the expertise of the alliance's members, which includes paediatric and adult cardiac surgeons and cardiologists from all major units in Australia and New Zealand, as well as consumers and other key stakeholders.</p> <p>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.</p> <p>The CHAANZ registry and its ability to work with other relevant registries is the key resource required to facilitate the research priorities, enabling research collaboration, leveraging investment in the sector, and empowering patient and family interactions with the registry and its outcomes.</p> <p>Key deliverables:</p> <ol style="list-style-type: none"> <li>1. Integrate new information platforms with clinical practice to optimise transition of care to allow whole-of-life management in the environment where young adults with CHD will have the best outcomes.</li> <li>2. Document variations in care and to allow optimal systems planning to build capacity in health services.</li> <li>3. Facilitate research to understand Australian-relevant epidemiology and to determine effective interventions, and to identify relevant sub-groups of individuals relevant to recruitment to clinical trials for novel interventions.</li> </ol>

## RESEARCH – FOCUS AREA 6: RESEARCH PRIORITIES

ACTION	DETAIL
<p>6.5 Explore the role of exercise and increased activity in reducing the impact of CHD</p>	<p>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. Recent evidence from internationally-recognised Australian researchers has identified the benefits of specific types of exercise and indicated the potential benefits of a regular exercise training program as part of cardiac rehabilitation.</p> <p>At present, however, there is no clear consensus on the safety and effectiveness of such training. Targeted research on understanding the beneficial mechanisms of exercise on physiology can be rapidly applied to developing and testing relevant and personalised exercise regimes, leveraging the competitive advantage of Australian researchers to lead this field on the international stage.</p> <p>The opportunity for Australian researchers to collaborate with specialist accredited exercise physiologists to design and test appropriate exercise programs will also build the capacity to provide such cardiac rehabilitation interventions to CHD patients nationally in a cost-effective manner.</p> <p>Key deliverables:</p> <ol style="list-style-type: none"> <li>1. Determine the benefits of tailored exercise and activity programs on improved exercise tolerance, muscle strength, activity levels and quality of life.</li> <li>2. Define the mechanisms of how exercise and increases in activity benefit heart and physiological functions and adaptations that occur after cardiac rehabilitation.</li> <li>3. Design and test models for sustainability and compliance of exercise programs.</li> <li>4. Develop guidelines for personalised exercise and activity programs relevant to the large range of structural variations in CHD, in coordination with CSANZ.</li> <li>5. Develop professional education in collaboration with Exercise and Sports Science Australia (ESSA) to build capacity of specialist cardiac-accredited exercise physiologists, available to provide tailored programs to CHD patients nationally.</li> </ol>

## FOCUS AREA 7: SURVEILLANCE

### DESIRED OUTCOMES

Australia and New Zealand will have a CHD registry which captures every baby born with CHD and tracks their CHD journey to inform clinical outcomes and provide the best whole of life care.

Australia will understand for the first time the hidden costs of CHD in the health system, to the government and families impacted by CHD, in order to identify opportunities to transform care, economic and financial burden.

### WHY?

**Surveillance** is the ongoing, systematic collection, analysis and interpretation of health-related data essential to the planning, implementation, and evaluation of public health practice.<sup>152</sup>

Effective surveillance of CHD is needed to understand and monitor the epidemiology of the disease to set priorities and guide health policy and strategies.

As recommended in this Action Plan, a CHD taskforce should be established. The CHD taskforce would play a role in ensuring the surveillance, monitoring and evaluation of the Action Plan.

Surveillance will include:

- use the CHAANZ registry and national survey of CHD patients to track the impact of Action Plan recommendations over time;
- conduct focus group interviews of key stakeholders and implementation partners;
- undertake three economic impact reports in years one, three and five (post report release) to measure any improvements to the economic and productivity burden of the disease on the Australian community; and
- publish an Action Plan annual report card (Action 7.3).

## RESEARCH – FOCUS AREA 7: SURVEILLANCE

ACTION	DETAIL
7.1 Ongoing investment in the Congenital Heart Alliance of Australia and New Zealand (CHAANZ) Registry	Accelerated implementation of the CHAANZ Bi-National Congenital Heart Disease Registry, and harmonisation of several existing smaller condition specific registries.
7.2 Invest in an economic burden of CHD report	It is proposed as an adjunct to the Registry to engage a health economist to prepare a consensus model for measurement of the economic burden of CHD (both direct and indirect costs) and to update this report every two years to monitor the impact of the Action Plan.
7.3 Invest in funding of the annual national CHD survey	Continue funding an annual national CHD survey to collect data from people with CHD, including their healthcare use and experiences, and the health, social and economic impact of living with CHD.
7.4 Develop an evaluation framework that includes production of an annual report card to monitor progress against this Action Plan	HeartKids, in conjunction with the CHD taskforce and key stakeholders, will develop an evaluation framework to monitor the Action Plan progress and to generate an annual report card. Key inputs would include the CHD registry and survey, stakeholder focus groups, economic burden of disease reports and likely external data sources, for example AIHW.

## ACKNOWLEDGEMENTS

Development of the Action Plan was led by HeartKids with funding from the Australian Government Department of Health.

Development of the Action Plan took place during 2018. Many individuals and organisations contributed time and expertise to the development of the Action Plan, including people with CHD, health professionals, key medical and cardiac organisations, the research community and health departments within the Australian, state and territory governments.

HeartKids sincerely thanks the members of the Steering Committee and Expert Working Groups, including the hardworking chairs, and all those who participated in the extensive consultation and development phase. The involvement and willingness of all concerned to share their experience and expertise in order to improve outcomes for people with CHD is greatly appreciated. Members of the Steering Committee are listed below. Members of each of the five Expert Working Group are listed in Compendium A – Consultation Summary and Implementation Partners.

Steering Committee members:

- Ms Jan McClelland AM (Chair), Chair of the Board of HeartKids
- Professor David Celermajer, CHAANZ Steering Committee; Board Director, HeartKids; Royal Prince Alfred Hospital; University of Sydney
- Associate Professor Michael Cheung, Director of Cardiology, Royal Children’s Hospital Melbourne
- Professor Yves d’Udekem, Deputy Director, Department of Cardiac Surgery, Royal Children’s Hospital Melbourne
- Dr Andrew Kelly, Medical Unit Head Cardiology, Women’s and Children’s Hospital Adelaide
- Associate Professor Gary Sholler, Director Cardiac Services, Sydney Children’s Hospital Network
- Associate Professor Robert Justo, Director Paediatric Cardiology, Queensland Health
- Dr James Ramsay, Paediatric Cardiologist, Perth Children’s Hospital
- Dr Gavin Wheaton, Medical Director, Women’s and Children’s Hospital Adelaide
- Dr Bo Remenyi, Paediatric Cardiologist, Royal Darwin Hospital; Menzies School of Health
- Dr Rosemary Wyber, Head of Strategy END RHD, Telethon Kid’s Institute
- Mr Mark Brooke, Chief Executive Officer, HeartKids (until September 2018)
- Dr Lisa Selbie, Chair, Research Advisory Committee, Board Director, HeartKids
- Ms Elizabeth Flynn, Acting First Assistant Secretary, Population Health and Sport, Department of Health
- Ms Jacinta McDonald, Director Chronic Disease Policy Section, Department of Health
- Ms Michelle Pendrick, consumer representative, adult with CHD
- Ms Simone Wright, consumer representative, parent of a CHD child.

## APPENDIX 1: FRAMEWORK FOR ACTION

Priorities, focus areas, recommended actions and outcomes of the Action Plan.

PRIORITY	FOCUS AREA	RECOMMENDED ACTIONS	OUTCOMES
MANAGEMENT, CARE AND SUPPORT	1. STANDARDS OF CARE	1.1 Develop national standards of care for CHD	Australia will have its first national CHD standards of care, including world first neurodevelopmental and mental healthcare standards.
		1.2 Develop authoritative national clinical practice guidelines on CHD for health services and health practitioners throughout Australia	
		1.3 Establish a CHD taskforce to oversee the development of standards of care for CHD	
	2. INFRASTRUCTURE	2.1 Develop a CHD infrastructure plan	Australia will have a health system infrastructure which provides continuity of specialised care for CHD patients from birth through adolescence to adulthood.
		2.2 Invest in technological infrastructure	
		2.3 Deliver education, training and support for health professionals	
		2.4 Support the management of CHD in primary care	
		2.5 Disseminate best practice information and approaches throughout services in Australia	
3. AWARENESS AND EDUCATION	3.1 Increase access to quality, relevant information, education and support through a range of approaches including phone, face to face and digital	People with CHD and their families will have access to information, support and resources when and where they are needed.	
	3.2 Increase access to ongoing support by funding inpatient and outpatient community (peer) support team members		

PRIORITY	FOCUS AREA	RECOMMENDED ACTIONS	OUTCOMES
SUPPORTIVE COMMUNITIES		3.3 Increase the number of young people engaged with transition and ongoing specialist CHD care	
		3.4 Deliver awareness and education for non-health professionals	
	4. NEURO DEVELOPMENTAL AND MENTAL HEALTHCARE	4.1 Develop standards of mental healthcare for CHD	People with CHD and their families will experience improved mental health, cognitive outcomes and quality of life as a result of world first neurodevelopmental and mental health standards.
		4.2 Develop standards of neurodevelopmental care for CHD	
		4.3 Establish clear models of care	
	5. PRIORITY POPULATIONS	5.1 Fund CHD cardiac coordinators in each jurisdiction	Australia will have a health system that is responsive to the specific needs of identified priority populations with CHD through provision of infrastructure and continuity of specialised care from birth through adolescence to adulthood.
		5.2 Fund cardiac sonographers and a portable echo machine in each jurisdiction	
		5.3 Evaluate current specialist outreach services for regional and remote communities, including remote Aboriginal communities, and fund outreach services where significant gaps exist	
		5.4 Establish models of care for the transition of care from paediatric to adult cardiac health services, including investing in transition nurses	
		5.5 Investigate options for reducing barriers to accessing care and supporting people with CHD and their families	

PRIORITY	FOCUS AREA	RECOMMENDED ACTIONS	OUTCOMES
RESEARCH	6. RESEARCH PRIORITIES	6.1 Understand the genetic causes of and predisposition to CHD to provide support for those impacted by CHD	<p>Australia will lead the way in research to:</p> <ul style="list-style-type: none"> <li>identify the factors that cause CHD, both genetic and environmental, to work towards reducing prevalence in the future;</li> <li>identify the burden of neurodevelopmental, psychosocial, poor mental health and economic factors on patients with CHD in order to improve the quality of life of CHD patients and their families;</li> <li>identify the health factors that impact CHD pre- and post-natally to inform evidence base and transform care;</li> <li>understand Australian-relevant epidemiology to determine effective interventions; and</li> <li>inform the development of an evidence-supported exercise prescription for patients with CHD.</li> </ul>
		6.2 Prevent neurodevelopmental and mental health complications	
		6.3 Understand the impact of prenatal and postnatal factors affecting the health of children born with CHD and their families	
		6.4 Measure longitudinal impact and inform practice with the National Congenital Heart Disease Registry	
		6.5 Explore the role of exercise and increased activity in reducing the impact of CHD	
	7. SURVEILLANCE	7.1 Ongoing investment in the Congenital Heart Alliance of Australia and New Zealand (CHAANZ) Registry	Australia and New Zealand will have a CHD registry which captures every baby born with CHD and tracks their CHD journey to inform clinical outcomes and provide the best whole of life care.
		7.2 Invest in an economic burden of CHD report	Australia will understand for the first time the hidden costs of CHD in the health system, to the government and families impacted by CHD, in order to identify opportunities to transform care, economic and financial burden.
		7.3 Invest in funding of the annual national CHD survey	
		7.4 Develop an evaluation framework that includes production of an annual report card to monitor progress against this Action Plan	

## APPENDIX 2: GLOSSARY

**Access to care** means having the timely use of healthcare services to achieve the best health outcomes.

**Awareness and education** embrace the range of evidence-based communications, information and learning approaches often used in health promotion, health education and health literacy to bring about positive change in public health. These approaches often incorporate organised comprehensive communications activities (including campaigns, response services and resources) designed to raise awareness, motivate an attitude or behaviour change and improve outcomes for individuals and populations.

**Cardiac sonographers** are part of the multidisciplinary team that cares for people with CHD. Cardiac sonographers (scientific staff dedicated to ultrasound imaging) are specially trained to perform echocardiograms, or ultrasound imaging, to evaluate different aspects of the heart and help doctors diagnose and treat patients with heart and blood vessel conditions.

**Care continuum** is a concept involving an integrated system of care that guides and tracks a patient over time through a comprehensive array of health services.

**Childhood heart disease** (CHD) is a general term for a range of conditions that affect the normal workings of the heart. CHD can be present at birth (congenital) or be acquired or appear for the first time after birth. CHD is a chronic condition that, for many patients requires complex, specialised care across the life course.

**CHD cardiac coordinators** are proposed as a new and core part of the multidisciplinary team that cares for people with CHD. CHD cardiac coordinators will coordinate, and case-manage regional, rural and remote patients and families affected by CHD, with a focus on Aboriginal and Torres Strait Islander people and adolescent/young adult patients.

**CHD Research** is research into the causes, treatment and management of CHD and involves medical and health research.<sup>153</sup>

**Chronic conditions** are often used interchangeably with ‘chronic diseases’, ‘noncommunicable diseases’, and ‘long-term health conditions’. Chronic conditions have complex and multiple causes; may affect individuals either alone or as co-morbidities; usually have a gradual onset, although they can have sudden onset and acute stages; occur across the lifecycle, although they become more prevalent with older age; can compromise quality of life and create limitations and disability; are long-term and persistent, and often lead to a gradual deterioration of health and loss of independence. While not usually life-threatening, are the most common and leading cause of premature mortality.<sup>154</sup>

**Clinical quality registries (CQR)** are organisations which systematically monitor the quality (appropriateness and effectiveness) of healthcare within specific clinical domains, by routinely collecting, analysing and reporting health-related information.<sup>155</sup> Clinical trials are research investigations in which people volunteer to test new treatments, interventions or tests as a means to prevent, detect, treat or manage various diseases or medical conditions.<sup>156</sup>

**Co-morbidity** describes two or more disorders, conditions or illnesses occurring in the same person. They can occur at the same time or one after the other. Co-morbidity also implies interactions between the illnesses that can worsen the course of both.

**Consumer engagement** in research means consumers, community members, researchers and research organisations working in partnerships, to improve the health and wellbeing of all Australians through health and medical research.<sup>157</sup>

**Culturally appropriate services** are broadly defined as care and services that are respectful of and responsive to the cultural and linguistic needs of all individuals.

**A disability** is any condition of the body or mind (impairment) that makes it more difficult for the person with the condition to do certain activities and interact with the world around them.



**Disparities** refer to great differences in health outcomes between populations. Race or ethnicity, sex, sexual identity, age, disability, socioeconomic status, and geographic location all contribute to an individual's ability to achieve good health.

An **echocardiogram**, often referred to as a cardiac echo or simply an echo, is a specialised ultrasound examination of the heart. Echocardiograms are performed by cardiac sonographers.

**Echocardiography** uses standard two-dimensional, three-dimensional, and Doppler ultrasound to create images of the heart. Echocardiography should not be confused with an ECG (abbreviation for electrocardiogram), which is a recording of the electrical activity of the heart.

**Evidence-based practice(s)** is the conscientious, explicit, and judicious use of current best evidence when making decisions about the care of patients with CHD. This involves integrating individual clinical expertise with the best available external clinical evidence from systematic research.

**Fetus/foetal heart disease is heart disease**, such as CHD, which is recognised before birth, resulting in patients requiring a range of unique management plans and services plans.

**Fontan** The Fontan circulation is a type of reconstruction of the heart and circulation applied to patients with a complex cardiac condition characterised by a variety of forms of single ventricle (single heart pump in contrast to the usual two fully-formed pumps). The patients have a very specific set of complex early and late care requirements.

**Genetics** is the study of heredity.<sup>158</sup>

**Genomics** is the study of genes and their functions, and related techniques.<sup>159</sup>

**Healthcare cost:** direct vs. indirect costs are those costs borne by the healthcare system, community, and patients' families in addressing an illness. Indirect costs are mainly productivity losses to society caused by the health problem or disease.

**Health determinants** are factors which influence health. Many factors combine to affect the health of individuals and communities. The determinants of health include the social and economic environment, the physical environment, and the person's individual characteristics and behaviours.

**Health literacy** is the degree to which individuals have the capacity to obtain, process, and understand basic health information and services needed to make appropriate health decisions.

'**Heart kids**' are children and adults living with CHD.

**Infrastructure** refers to the service and resource requirements needed to deliver quality care for people with CHD who have complex health needs and require lifelong care from workforce operating from a range of institutions, both public and private.

**Loss to follow-up/lost to care** refers to patients who were at one point actively participating in cardiac care services but have become lost (either by error in a computer tracking system or by being unreachable) at the point of follow-up. Over 50 per cent of Australians with CHD are being lost to care when transitioning from paediatric to adult cardiac health services.

**Mental healthcare for CHD** is defined as integrated, specialised and evidence-based care that focuses on reducing mental health burden and optimising emotional and psychobiological wellbeing for affected foetuses, infants, children, young people and adults and their families. There is an emphasis on prevention and early intervention through evidence-based screening, assessment and psychological care and treatment, as part of routine cardiac care.

**Mortality rate** is a measure of the frequency of occurrence of death in a defined population during a specified interval.

**A multidisciplinary team**, often abbreviated as MDT, involves a range of healthcare professionals working together to deliver comprehensive patient care. Collaborative teams vary according to patients' needs, patient load, organisational constraints, resources, clinical setting, geographic location, and professional skills.

**Neonate** is a newborn baby in the first 28 days of life. In some cases of premature infants, neonatal care will continue until the equivalence of term gestation or weight is reached.

**Neurodevelopmental care** is defined as integrated, specialised and evidence-based care to prevent or minimise neurodevelopmental or neurocognitive impairment in infants, children, young people and adults with CHD, with an emphasis on early intervention, ongoing surveillance and specialised intervention, as indicated.

**Patient registries** are organised systems that use observational study methods to collect uniform data (clinical and other) to evaluate specified outcomes for a population defined by a particular disease, condition, or exposure and that serves predetermined scientific, clinical, or policy purpose(s).

**Peer support occurs** when people provide knowledge, experience, emotional, social or practical assistance to each other. Peer support is distinct from other forms of social support in that the source of support is a peer; a peer is in a position to offer support by virtue of relevant, lived experience.

**Person-centred and family-centred care** is defined as healthcare that establishes a partnership among practitioners, patients, and their families (when appropriate) to ensure that healthcare decisions are respectful of and responsive to the preferences, needs and values of patients, and that they have the education and support they need to make decisions and participate in their own care.

**Precision medicine** is an emerging approach for disease prevention and management that tailors care to account for an individual's variations in genes, environment, and lifestyle.

**Prevalence** is the proportion of a population which has (or had) a specific characteristic in a given time period. In medicine, this is typically an illness, a condition, or a risk factor, such as depression or smoking.

**Prevention** involves action at the primary, secondary and tertiary levels to prevent disease. In the case of CHD, primary prevention strategies that aim to limit the incidence of disease and disability in the population are focused on preventing the onset of rheumatic heart disease (RHD) which is entirely preventable, particularly among Aboriginal and Torres Strait Islander people. There is no evidence for effective interventions to prevent congenital heart disease, therefore congenital heart disease is targeted with secondary prevention (early detection/intervention) and tertiary prevention (reducing the progression of the disease and improving the individual's quality of life) strategies, including in neurodevelopmental and mental healthcare.<sup>160</sup>

**Primary healthcare** for most patients is a primary care clinician who will be their first point of contact in the Australian health system. A primary care clinician may be a doctor, dentist, nurse, allied health professional or a pharmacist. This level of care may be provided in a general practice, community or allied health centre or Aboriginal and Community Controlled Health Services. It may also include health promotion, health education or prevention. Depending on the person's health condition, they may be referred on to secondary or tertiary care.

**Priority populations** are population groups that are identified as a priority target audience for action to prevent and manage chronic conditions. Some populations are disproportionately affected due to a complex interaction between the physical environment, social and cultural determinants and biomedical and behavioural risk factors. This is demonstrated by a higher prevalence of CHD and a greater burden of disease, resulting in inequitable health outcomes.<sup>161</sup> Populations may be prioritised for a number of additional reasons, including, for example, facing unique challenges with their condition, barriers to accessing care, or an increased risk due to the presence of risk factors.

**Research impact** is the contribution that research makes to the economy, society, environment or culture, beyond the contribution to academic research.<sup>162</sup>



**Rheumatic heart disease (RHD)** is chronic damage to the valves of the heart, caused by repeated episodes of acute rheumatic fever (ARF). ARF is caused by an auto-immune reaction to an infection with the bacterium group A streptococcus (GAS).<sup>163</sup>

**Secondary healthcare** refers to health services provided by medical specialists and other health professionals who do not have the first contact with patients. Secondary care is usually (but not always) delivered in a hospital/clinic with the initial referral being made by the primary care professional. Secondary healthcare can also refer to ongoing services not necessarily provided in the hospital, such as psychiatrists, physiotherapists and occupational therapists.<sup>164</sup>

**Self-management** can be defined as the decisions and behaviours that people with CHD engage in that affect their health. Self-management support is the care and encouragement provided to people with CHD and their families to help them understand their central role in managing their illness, make informed decisions about care, and engage in healthy behaviours. Self-management support can assist and empower people to learn more about their conditions and to take an active role in their healthcare.

**Standards of care** are components of care that health professionals and health services should offer patients for a specific clinical condition. Standards define and uphold the safety and quality of healthcare provision and provide a framework to ensure people receive the best possible care and outcomes.<sup>165</sup> They guide delivery of care and the related service and resource requirements. Standards of care are best implemented on a national basis for areas of special need.

**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.<sup>166</sup>

**Telehealth** allows healthcare professionals to examine, diagnose, and treat patients using technology like a phone, computer, or other connected device.

**Tertiary healthcare** refers to highly-specialised healthcare, often for inpatients and on referral from a primary or secondary health professional. This often includes particularly complex medical or surgical procedures.<sup>167</sup>

**Transition** refers to the transition of care from paediatric to adult health services. Transition is a coordinated process involving the young person, their families or carers and all hospital departments involved in their care. It is a gradual process, beginning in early adolescence and is not completed until the young person has engaged in an adult health service. A holistic transition approach focuses on empowering young people and their parents/carers by advancing their knowledge of medical needs and providing support and knowledge (and additional care pathways if relevant) in the areas of psychosocial and psychological wellbeing; educational and vocational needs; and assistance with community linkages and support.<sup>168</sup>

## REFERENCES

- <sup>1</sup> CELERMAJER, D., STRANGE, G., CORDINA, R., SELBIE, L., SHOLLER, G., WINLAW, D., ALPHONSO, N., JUSTO, R., NICHOLAE, M., KASPARIAN, N., WEINTRAUB, R. G., CHEUNG, M., GRIGG, L. E., BRIZARD, C. P., WHEATON, G., DISNEY, P., STEWART, S., BULLOCK, A., RAMSAY, J., GENTLES, T. & D'UDEKEM, Y. (2016). "Congenital Heart Disease Requires a Lifetime Continuum of Care: A call for a Regional Registry." *Heart, Lung and Circulation* **25**(8): 750-754.
- <sup>2</sup> AUSTRALIAN INSTITUTE OF HEALTH AND WELFARE (2017). "Heart, stroke & vascular diseases." Retrieved June 2018, from <https://www.aihw.gov.au/reports-data/health-conditions-disability-deaths/heart-stroke-vascular-diseases/overview>.
- <sup>3</sup> AUSTRALIAN INSTITUTE OF HEALTH AND WELFARE (2018). "Deaths in Australia." Retrieved June 2018, from <https://www.aihw.gov.au/reports/life-expectancy-death/deaths-in-australia/contents/life-expectancy>.
- <sup>4</sup> DAVIES, S., HOFER, A. & REEVE, C. (2014). "Mortality attributable to rheumatic heart disease in the Kimberley: a data linkage approach." *Internal Medicine Journal* **44**: 1074-1080.
- <sup>5</sup> CELERMAJER, D., STRANGE, G., CORDINA, R., SELBIE, L., SHOLLER, G., WINLAW, D., ALPHONSO, N., JUSTO, R., NICHOLAE, M., KASPARIAN, N., WEINTRAUB, R. G., CHEUNG, M., GRIGG, L. E., BRIZARD, C. P., WHEATON, G., DISNEY, P., STEWART, S., BULLOCK, A., RAMSAY, J., GENTLES, T. & D'UDEKEM, Y. (2016). "Congenital Heart Disease Requires a Lifetime Continuum of Care: A call for a Regional Registry." *Heart, Lung and Circulation* **25**(8): 750-754.
- <sup>6</sup> KHAIRY, P., IONESCU-ITTU, R., MACKIE, A. S., ABRAHAMOWICZ, M., PILOTE, L. & MARELLI, A. (2010). "Changing mortality in congenital heart disease." *Journal of the American College of Cardiology* **56**: 1149-1157.
- <sup>7</sup> LEGGAT, S. (2011). The 2011 Childhood Heart Disease in Australia White Paper. Childhood heart disease in Australia: Current Practices and Future Needs. A report for HeartKids and Paediatric and Congenital Council of the Cardiac Society of Australia and New Zealand. Pennant Hills, NSW, Australia, HeartKids Australia.
- <sup>8</sup> LE GLOAN, L., MERCIER, L., DORE, A., MARCOTTE, F., IBRAHIM, R., MONGEON, F., ASGAR, A., MIRO, J., POIRIER, N. & KHAIRY, P. (2011). "Recent advances in adult congenital heart disease." *Circulation* **75**: 2287-2295.
- <sup>9</sup> WINLAW, D. (2007). "Congenital heart disease in the 21st century." *Critical Care and Resuscitation* **9**: 270-274.
- <sup>10</sup> LEGGAT, S. (2011). The 2011 Childhood Heart Disease in Australia White Paper. Childhood heart disease in Australia: Current Practices and Future Needs. A report for HeartKids and Paediatric and Congenital Council of the Cardiac Society of Australia and New Zealand. Pennant Hills, NSW, Australia, HeartKids Australia.
- <sup>11</sup> AUSTRALIAN HEALTH MINISTERS' ADVISORY COUNCIL (2017). National Strategic Framework for Chronic Conditions. Canberra, ACT, Australia, Australian Government.
- <sup>12</sup> VICTORIA STATE GOVERNMENT (2014). "Heart abnormality birth defects." Retrieved February 2018, from <https://www.betterhealth.vic.gov.au/health/ConditionsAndTreatments/heart-abnormality-birth-defects?viewAsPdf=true>.
- <sup>13</sup> CELERMAJER, D., STRANGE, G., CORDINA, R., SELBIE, L., SHOLLER, G., WINLAW, D., ALPHONSO, N., JUSTO, R., NICHOLAE, M., KASPARIAN, N., WEINTRAUB, R. G., CHEUNG, M., GRIGG, L. E., BRIZARD, C. P., WHEATON, G., DISNEY, P., STEWART, S., BULLOCK, A., RAMSAY, J., GENTLES, T. & D'UDEKEM, Y. (2016). "Congenital Heart Disease Requires a Lifetime Continuum of Care: A call for a Regional Registry." *Heart, Lung and Circulation* **25**(8): 750-754.

---

<sup>14</sup> AUSTRALIAN INSTITUTE OF HEALTH AND WELFARE (2017). "Heart, stroke & vascular diseases." Retrieved June 2018, from <https://www.aihw.gov.au/reports-data/health-conditions-disability-deaths/heart-stroke-vascular-diseases/overview>.

<sup>15</sup> AUSTRALIAN INSTITUTE OF HEALTH AND WELFARE (2018). "Deaths in Australia." Retrieved June 2018, from <https://www.aihw.gov.au/reports/life-expectancy-death/deaths-in-australia/contents/life-expectancy>.

<sup>16</sup> CELERMAJER, D., STRANGE, G., CORDINA, R., SELBIE, L., SHOLLER, G., WINLAW, D., ALPHONSO, N., JUSTO, R., NICHOLAE, M., KASPARIAN, N., WEINTRAUB, R. G., CHEUNG, M., GRIGG, L. E., BRIZARD, C. P., WHEATON, G., DISNEY, P., STEWART, S., BULLOCK, A., RAMSAY, J., GENTLES, T. & D'UDEKEM, Y. (2016). "Congenital Heart Disease Requires a Lifetime Continuum of Care: A call for a Regional Registry." *Heart, Lung and Circulation* **25**(8): 750-754.

<sup>17</sup> VICTORIA STATE GOVERNMENT (2014). "Heart abnormality birth defects." Retrieved February 2018, from <https://www.betterhealth.vic.gov.au/health/ConditionsAndTreatments/heart-abnormality-birth-defects?viewAsPdf=true>.

<sup>18</sup> Ibid.

<sup>19</sup> CELERMAJER, D., STRANGE, G., CORDINA, R., SELBIE, L., SHOLLER, G., WINLAW, D., ALPHONSO, N., JUSTO, R., NICHOLAE, M., KASPARIAN, N., WEINTRAUB, R. G., CHEUNG, M., GRIGG, L. E., BRIZARD, C. P., WHEATON, G., DISNEY, P., STEWART, S., BULLOCK, A., RAMSAY, J., GENTLES, T. & D'UDEKEM, Y. (2016). "Congenital Heart Disease Requires a Lifetime Continuum of Care: A call for a Regional Registry." *Heart, Lung and Circulation* **25**(8): 750-754.

<sup>20</sup> VICTORIA STATE GOVERNMENT (2014). "Heart abnormality birth defects." Retrieved February 2018, from <https://www.betterhealth.vic.gov.au/health/ConditionsAndTreatments/heart-abnormality-birth-defects?viewAsPdf=true>.

<sup>21</sup> ALANKARAGE, D., IP, E., SZOT, J., MUNRO, J., BLUE, G., HARRISON, K., CUNY, H., ENRIQUEZ, A., TROUP, and H. M., D., WILSON, M., HARVEY, R., SHOLLER, G., GRAHAM, R., HO, J., KIRK, E., PACHTER, N., CHAPMAN, G., WINLAW, D., GIANNOULATOU, E. & DUNWOODIE, S. (2018). "Identification of clinically actionable variants from genome sequencing of families with congenital heart disease." from <https://www.nature.com/articles/s41436-018-0296-x>.

<sup>22</sup> Ibid.

<sup>23</sup> SHOLLER, G., KASPARIAN, N., PYE, V., COLE, A. & WINLAW, D. (2011). "Fetal and post-natal diagnosis of major congenital heart disease: Implications for medical and psychological care in the current era." *Journal of Paediatrics and Child Health* **47**: 711-722.

<sup>24</sup> BELLINGER, D., WYPIJ, D., DUPLESSIS, A., RAPPAPORT, L., JONAS, R., WERNOVSKY, G. & NEWBURGER, J. (2003). "Neurodevelopmental status at eight years in children with dextroposition of the great arteries: the Boston Circulatory Arrest Trial." *Journal of Thoracic and Cardiovascular Surgery* **126**: 1385-1396.

<sup>25</sup> DONOFRIO, M., et al. (2011). "Impact of congenital heart disease on fetal brain development and injury." *Current Opinion in Paediatrics* **23**: 502-511.

<sup>26</sup> DONOFRIO, M. M., A. (2010) Impact of Congenital Heart Disease on Brain Development and Neurodevelopmental Outcome. *International Journal of Paediatrics* DOI: doi: 10.1155/2010/359390.

<sup>27</sup> MAJNEMER, A., LIMPEROPOULOS, C., SHEVELL, M. & ROSENBLATT, B. (2006). "Long-term neuromotor outcome at school entry of infants with congenital heart defects requiring open-heart surgery." *Journal of Paediatrics* **148**: 72-77.

- 
- <sup>28</sup> MARINO, B., LIPKIN, P., NEWBURGER, J., PEACOCK, G., GERDES, M., GAYNOR, J., MUSSATTO, K., UZARK, K., GOLDBERG, C., JOHNSON WH JR, LI, J., SMITH, S., BELLINGER, D. & MAHLE, W. (2012). "Neurodevelopmental outcomes in children with congenital heart disease: evaluation and management: a scientific statement from the American Heart Association." *Circulation* **126**: 1143-1172.
- <sup>29</sup> KASPARIAN, N., WINLAW, D. & SHOLLER, G. (2016). "'Congenital heart health': how psychological care can make a difference." *The Medical Journal of Australia* **205**: 104-107.
- <sup>30</sup> VICTORIA STATE GOVERNMENT (2014). "Heart abnormality birth defects." Retrieved February 2018, from <https://www.betterhealth.vic.gov.au/health/ConditionsAndTreatments/heart-abnormality-birth-defects?viewAsPdf=true>.
- <sup>31</sup> RHDAUSTRALIA (2012). Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition). Quick reference guides: RHD Australia (ARF/RHD writing group). National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand.
- <sup>32</sup> AUSTRALIAN INSTITUTE OF HEALTH AND WELFARE (2013). Rheumatic heart disease and acute rheumatic fever in Australia: 1996–2012. Cardiovascular disease series. Canberra, ACT, Australia, Australian Government.
- <sup>33</sup> WATKINS, D., JOHNSON, C., COLQUHOUN, S., KARTHIKEYAN, G., BEATON, A., BUKHMAN, G., FOROUZANFAR, H., LONGENECKER, C., MAYOSI, B., MENSAH, G., NASCIMENTO, B., RIBEIRO, A., SABLE, C., STEER, A., NAGHAVI, M., MOKDAD, A., MURRAY, C., VOS, T., CARAPETIS, J., & ROTH, G. (2017) Global, Regional, and National Burden of Rheumatic Heart Disease, 1990–2015. *The New England Journal of Medicine* **377**, 713-722 DOI: 10.1056/NEJMoa1603693.
- <sup>34</sup> REMENYI, B., CARAPETIS, J., WYBER, R., TRAUBERT, K. & MAYOSI, B. (2013) Position statement of the World Heart Federation on the prevention and control of rheumatic heart disease. *Nature Reviews Cardiology* **10**, 284-292 DOI: 10.1038/nrcardio.2013.34.
- <sup>35</sup> RHDAUSTRALIA (2012). Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition). Quick reference guides: RHD Australia (ARF/RHD writing group). National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand.
- <sup>36</sup> BHARUCHA, T., LEE, K.J., DAUBENEY, P.E., NUGENT, A.W., TURNER, C., SHOLLER, G.F. ROBERTSON, T., JUSTO, R., RAMSAY, J., CARLIN, J.B., COLAN, S.D., KING, I., WEINTRAUB, R.G., DAVIS, A.M. (2015). "NACCS (National Australian Childhood Cardiomyopathy Study) Investigators." *Journal American College of Cardiology* **65**(21): 2302-2310.
- <sup>37</sup> CANTER, C. E. and K. E. SIMPSON (2014). "Diagnosis and treatment of myocarditis in children in the current era." *Circulation* **129**: 115-128.
- <sup>38</sup> FOERSTER, S. and C. CANTER (2011). "Contemporary etiology, outcomes, and therapy in pediatric myocarditis." *Progress in Pediatric Cardiology* **31**: 123-128.
- <sup>39</sup> DIMITRIADES, V. R., BROWN, A.G. & GEDALIA, A. (2014). "Kawasaki disease: pathophysiology, clinical manifestations, and management." *Current Rheumatology Reports* **16**(6): 423.
- <sup>40</sup> MCCRINDLE, B. W., ROWLEY, A.H., NEWBURGER, J.W., BURNS, J.C., BOLGER, A.F., GEWITZ, M., BAKER, A.L., JACKSON, M.A., TAKAHASHI, M., SHAH, P.B., KOBAYASHI, T., WU, M.H., SAJI, T.T., PAHL, E., AMERICAN HEART ASSOCIATION RHEUMATIC FEVER, ENDOCARDITIS, AND KAWASAKI DISEASE COMMITTEE OF THE COUNCIL ON CARDIOVASCULAR DISEASE IN THE YOUNG, COUNCIL ON CARDIOVASCULAR AND STROKE NURSING, COUNCIL ON CARDIOVASCULAR SURGERY AND ANESTHESIA & COUNCIL ON EPIDEMIOLOGY AND PREVENTION. (2017). "Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals From the American Heart Association." *Circulation* **135**(17): e927-e999.

<sup>41</sup> SON, M. and J. NEWBURGER (2018) Kawasaki Disease. *Pediatrics in Review* **39**, 78-90 DOI: 10.1542/PIR.2016-0182.

<sup>42</sup> KATHIRESAN, S. and D. SRIVASTAVA (2012). "Genetics of Human Cardiovascular Disease." *Cell* **148**: 1242-1257.

<sup>43</sup> BAGNALL, R. D., WEINTRAUB, R.G., INGLES, J., DUFLOU, J., YEATES, L., LAM, L., DAVIS, A.M., THOMPSON, T., CONNELL, V., WALLACE, J., NAYLOR, C., CRAWFORD, J., LOVE, D.R., HALLAM, L., WHITE, J., LAWRENCE, C., LYNCH, M., MORGAN, N., JAMES, P., DU SART, D., PURANIK, R., LANGLOIS, N., VOHRA, J., WINSHIP, I., ATHERTON, J., MCGAUGHRAN, J., SKINNER, J.R. & SEMSARIAN, C. (2016). "A Prospective Study of Sudden Cardiac Death among Children and Young Adults." *New England Journal Medicine* **374**(25): 2441-2452.

<sup>44</sup> BRUGADA, J., BLOM, N., SARQUELLA-BRUGADA, G., BLOMSTROM-LUNDQVIST, C., DEANFIELD, J., JANOUSEK, J., ABRAMS, D., BAUERSFELD U., BRUGADA, R., DRAGO, DE GROOT, N., HAPPONEN, J.M., HEBE, J., YEN HO, S., MARIJON, E., PAUL, T., PFAMMATTER, J.P., ROSENTHAL, E., EUROPEAN HEART RHYTHM ASSOCIATION., ASSOCIATION FOR EUROPEAN PAEDIATRIC AND CONGENITAL CARDIOLOGY. (2013). "Pharmacological and non-pharmacological therapy for arrhythmias in the pediatric population: EHRA and AEP-Arrhythmia Working Group joint consensus statement." *Europace* **15**: 1337-1382.

<sup>45</sup> PAUL, T., ABRAMS, D., AZIZ, P.F., BLOM, N.A., CHEN, J., CHESSA, M., COMBES, N., DAGRES, N., DILLER, G., ERNST, S., GIAMBERTI, A., HEBE, J., JANOUSEK, J., KNEBEL, T., MORENO, J., PEINADO, R., OISON, L., ROSENTHAL, E., SKINNER, J.R., ZEPPELFELD, K., ESC SCIENTIFIC DOCUMENT GROUP. (2018). "Arrhythmias in congenital heart disease: a position paper of the European Heart Rhythm Association (EHRA), Association for European Paediatric and Congenital Cardiology (AEP), and the European Society of Cardiology (ESC) Working Group on Grown-up Congenital heart disease, endorsed by HRS, PACES, APHRS, and SOLAECE." *Europace* **20**: 1719-1753.

<sup>46</sup> HEALTH POLICY ANALYSIS (2017). "Evaluation of the Commonwealth Rheumatic Fever Strategy - Final report." from [http://www.health.gov.au/internet/main/publishing.nsf/content/B6D489A5E5EC39B2CA25812A0078A371/\\$File/Rheumatic%20Fever%20Strategy%20Evaluation%20Final%20Report%20-%20HPA.PDF](http://www.health.gov.au/internet/main/publishing.nsf/content/B6D489A5E5EC39B2CA25812A0078A371/$File/Rheumatic%20Fever%20Strategy%20Evaluation%20Final%20Report%20-%20HPA.PDF).

<sup>47</sup> CELERMAJER, D., STRANGE, G., CORDINA, R., SELBIE, L., SHOLLER, G., WINLAW, D., ALPHONSO, N., JUSTO, R., NICHOLAE, M., KASPARIAN, N., WEINTRAUB, R. G., CHEUNG, M., GRIGG, L. E., BRIZARD, C. P., WHEATON, G., DISNEY, P., STEWART, S., BULLOCK, A., RAMSAY, J., GENTLES, T. & D'UDEKEM, Y. (2016). "Congenital Heart Disease Requires a Lifetime Continuum of Care: A call for a Regional Registry." *Heart, Lung and Circulation* **25**(8): 750-754.

<sup>48</sup> Ibid.

<sup>49</sup> STRANGE, G., STEWART, S., FARTHING, M., KASPARIAN, N.A., SELBIE, L.A., O'DONNELL, C., AYER, J., CORDINA, R. & CELERMAJER, D. (2018). Living with and caring for congenital heart disease in Australia: Insights from the congenital heart alliance of Australia and New Zealand online survey. *Heart, Lung and Circulation*.

<sup>50</sup> SECKELER, M., THOMAS, I., ANDREWS, J., JOINER, K. & KLEWER, S. (2016). "A review of the economics of adult congenital heart disease." *Expert Review of Pharmacoeconomics & Outcomes Research* **12**: 85-96.

<sup>51</sup> GALKAL (2018). Project Unite – A national picture of conditions and charities. A report for HeartKids. Sydney, NSW, Australia, GALKAL Independent Australian Strategy and Research Company.

<sup>52</sup> CELERMAJER, D., STRANGE, G., CORDINA, R., SELBIE, L., SHOLLER, G., WINLAW, D., ALPHONSO, N., JUSTO, R., NICHOLAE, M., KASPARIAN, N., WEINTRAUB, R. G., CHEUNG, M., GRIGG, L. E., BRIZARD, C. P., WHEATON, G., DISNEY, P., STEWART, S., BULLOCK, A., RAMSAY, J., GENTLES, T. & D'UDEKEM, Y. (2016). "Congenital Heart Disease Requires a Lifetime Continuum of Care: A call for a Regional Registry." *Heart, Lung and Circulation* **25**(8): 750-754.

- 
- <sup>53</sup> VAN DER LINDE, D., KONINGS, E., SLAGER, M., WITSENBURG, M., HELBING, W., TAKKENBERG, and J. J. & ROOS-HESELINK (2011). "Birth prevalence of congenital heart disease world-wide: a systematic review and meta-analysis." Journal of the American College of Cardiology **58**: 2241-2247.
- <sup>54</sup> AUSTRALIAN INSTITUTE OF HEALTH AND WELFARE (2017). "Heart, stroke & vascular diseases." Retrieved June 2018, from <https://www.aihw.gov.au/reports-data/health-conditions-disability-deaths/heart-stroke-vascular-diseases/overview>.
- <sup>55</sup> AUSTRALIAN INSTITUTE OF HEALTH AND WELFARE (2018). "Deaths in Australia." Retrieved June 2018, from <https://www.aihw.gov.au/reports/life-expectancy-death/deaths-in-australia/contents/life-expectancy>.
- <sup>56</sup> DAVIES, S., HOFER, A. & REEVE, C. (2014). "Mortality attributable to rheumatic heart disease in the Kimberley: a data linkage approach." Internal Medicine Journal **44**: 1074-1080.
- <sup>57</sup> LEGGAT, S. (2011). The 2011 Childhood Heart Disease in Australia White Paper. Childhood heart disease in Australia: Current Practices and Future Needs. A report for HeartKids and Paediatric and Congenital Council of the Cardiac Society of Australia and New Zealand. Pennant Hills, NSW, Australia, HeartKids Australia.
- <sup>58</sup> AUSTRALIAN INSTITUTE OF HEALTH AND WELFARE (2013). Rheumatic heart disease and acute rheumatic fever in Australia: 1996–2012. Cardiovascular disease series. Canberra, ACT, Australia, Australian Government.
- <sup>59</sup> AUSTRALIAN INSTITUTE OF HEALTH AND WELFARE (2017). Aboriginal and Torres Strait Islander health performance framework 2017: supplementary online tables. Canberra, ACT, Australia, Australian Institute of Health and Welfare.
- <sup>60</sup> DAVIES, S., HOFER, A. & REEVE, C. (2014). "Mortality attributable to rheumatic heart disease in the Kimberley: a data linkage approach." Internal Medicine Journal **44**: 1074-1080.
- <sup>61</sup> CHONG, L., FITZGERALD, D., CRAIG, J., MANERA, K., HANSON, C., CELERMAJER, D., AYER, J., KASPARIAN, N. & TONG, A. (2017). "Children's experiences of congenital heart disease: a systematic review of qualitative studies." European Journal of Pediatrics **177**: 319-336.
- <sup>62</sup> INGLES, J., SARINA, T., KASPARIAN, N. & SEMSARIAN, C. (2013). "Psychological wellbeing and posttraumatic stress associated with implantable cardioverter defibrillator therapy in young adults with genetic heart disease." International Journal of Cardiology **168**: 3779-3784.
- <sup>63</sup> KASPARIAN, N., WINLAW, D. & SHOLLER, G. (2016). "'Congenital heart health': how psychological care can make a difference." The Medical Journal of Australia **205**: 104-107.
- <sup>64</sup> KOVACS, A. H., SAIDI, A. S., KUHL, E. A., SEARS, S. F., SILVERSIDES, C., HARRISON, J. L., ONG, L., COLMAN, J., OECHSLIN, E. & NOLAN, R. P. (2009). "Depression and anxiety in adult congenital heart disease: predictors and prevalence." International Journal of Cardiology **137**: 158-164.
- <sup>65</sup> STEPTOE, A., MOHABIR, A., MAHON, N. & MCKENNA, W. (2000). "Health related quality of life and psychological wellbeing in patients with dilated cardiomyopathy." Heart **83**: 645-650.
- <sup>66</sup> WILSON, W., SMITH-PARRISH, M., MARINO, B. & KOVACS, A. (2015). "Neurodevelopmental and psychosocial outcomes across the congenital heart disease lifespan." Progress in Pediatric Cardiology **39**: 113-118.
- <sup>67</sup> Ibid.
- <sup>68</sup> CELERMAJER, D., STRANGE, G., CORDINA, R., SELBIE, L., SHOLLER, G., WINLAW, D., ALPHONSO, N., JUSTO, R., NICHOLAE, M., KASPARIAN, N., WEINTRAUB, R. G., CHEUNG, M., GRIGG, L. E., BRIZARD, C. P., WHEATON, G.,

---

DISNEY, P., STEWART, S., BULLOCK, A., RAMSAY, J., GENTLES, T. & D'UDEKEM, Y. (2016). "Congenital Heart Disease Requires a Lifetime Continuum of Care: A call for a Regional Registry." *Heart, Lung and Circulation* **25**(8): 750-754.

<sup>69</sup> CHAANZ (2018). Living with and caring for congenital heart disease in Australia: Insights from the congenital heart alliance of Australia and New Zealand online survey, Manuscript submitted for publication.

<sup>70</sup> LEE, W., FUNG, A., FONG, T. & YAN, B. (2016). "Long-Term Health-Related Economic Burden of Adult Congenital Heart Diseases in Hong Kong." *Value in Health* **19**: A865.

<sup>71</sup> RAJ, M., PAUL, M., SUDHAKAR, A., VARGHESE, A.A., HARIDAS, A.C., KABALI, C. & KUMAR, R.K. (2015). "Micro-Economic Impact of Congenital Heart Surgery: Results of a Prospective Study from a Limited-Resource Setting." *PLoS ONE* **10**(6): e0131348.

<sup>72</sup> GALKAL (2018). Project Unite – A national picture of conditions and charities. A report for HeartKids. Sydney, NSW, Australia, GALKAL Independent Australian Strategy and Research Company.

<sup>73</sup> KASPARIAN, N., WINLAW, D. & SHOLLER, G. (2017). "Childhood heart disease has a profound impact and is under-recognised. The Conversation." from <http://theconversation.com/childhood-heart-disease-has-a-profound-impact-and-is-under-recognised-84377>.

<sup>74</sup> KHAIRY, P., IONESCU-ITTU, R., MACKIE, A. S., ABRAHAMOWICZ, M., PILOTE, L. & MARELLI, A. (2010). "Changing mortality in congenital heart disease." *Journal of the American College of Cardiology* **56**: 1149-1157.

<sup>75</sup> LEGGAT, S. (2011). The 2011 Childhood Heart Disease in Australia White Paper. Childhood heart disease in Australia: Current Practices and Future Needs. A report for HeartKids and Paediatric and Congenital Council of the Cardiac Society of Australia and New Zealand. Pennant Hills, NSW, Australia, HeartKids Australia.

<sup>76</sup> LE GLOAN, L., MERCIER, L., DORE, A., MARCOTTE, F., IBRAHIM, R., MONGEON, F., ASGAR, A., MIRO, J., POIRIER, N. & KHAIRY, P. (2011). "Recent advances in adult congenital heart disease." *Circulation* **75**: 2287-2295.

<sup>77</sup> WINLAW, D. (2007). "Congenital heart disease in the 21st century." *Critical Care and Resuscitation* **9**: 270-274.

<sup>78</sup> CELERMAJER, D., STRANGE, G., CORDINA, R., SELBIE, L., SHOLLER, G., WINLAW, D., ALPHONSO, N., JUSTO, R., NICHOLAE, M., KASPARIAN, N., WEINTRAUB, R. G., CHEUNG, M., GRIGG, L. E., BRIZARD, C. P., WHEATON, G., DISNEY, P., STEWART, S., BULLOCK, A., RAMSAY, J., GENTLES, T. & D'UDEKEM, Y. (2016). "Congenital Heart Disease Requires a Lifetime Continuum of Care: A call for a Regional Registry." *Heart, Lung and Circulation* **25**(8): 750-754.

<sup>79</sup> SHOLLER, G., KASPARIAN, N., PYE, V., COLE, A. & WINLAW, D. (2011). "Fetal and post-natal diagnosis of major congenital heart disease: Implications for medical and psychological care in the current era." *Journal of Paediatrics and Child Health* **47**: 711-722.

<sup>80</sup> AUSTRALIAN HEALTH MINISTERS' ADVISORY COUNCIL (2017). National Strategic Framework for Chronic Conditions. Canberra, ACT, Australia, Australian Government.

<sup>81</sup> AUSTRALIAN COMMISSION ON SAFETY AND QUALITY IN HEALTHCARE (2013). "Clinical Care Standards Frequently asked questions (FAQs) for clinicians and health services." from <https://safetyandquality.gov.au/wp-content/uploads/2013/01/PDF-180KB.pdf>.

<sup>82</sup> CSANZ (2013). Adult Congenital Heart Disease (ACHD) Recommendations for Standards of Care., The Cardiac Society of Australia and New Zealand.

- 
- <sup>83</sup> NICOLAE, M., GENTLES, T., STRANGE, G., TANOUS, D., DISNEY, P., BULLOCK, A., GRIGG, L., CHEN, S.S.M., PRESSLEY, L., CORDINA, R., HORNUNG, T., O'DONNELL, C. & CELERMAJER, D.S. (in press). "Adult Congenital Heart Disease in Australia and New Zealand A Call for Optimal Care." [Heart, Lung and Circulation](#).
- <sup>84</sup> CORDINA, R., NASIR AHMAD, S., KOTCHETKOVA, I., EVEBORN, G., PRESSLEY, L., AYER, J., CHARD, R., TANOUS, D., ROBINSON, P., KILIAN, J., DEANFIELD, J. & CELERMAJER, D. (2018). "Management errors in adults with congenital heart disease: prevalence, sources, and consequences." [European Heart Journal](#) **39**: 982-989.
- <sup>85</sup> NHS ENGLAND (2016). Congenital Heart Disease Standards and Specifications. London, England, NHS England.
- <sup>86</sup> NHS SCOTLAND (2018). Specialist Congenital Heart Disease (CHD) standards for children and adults. S. C. C. SERVICES. Edinburgh, Scotland, Scottish Congenital Cardiac Service.
- <sup>87</sup> RHDAUSTRALIA (2012). Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition). Quick reference guides: RHD Australia (ARF/RHD writing group). National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand.
- <sup>88</sup> LEGGAT, S. (2011). The 2011 Childhood Heart Disease in Australia White Paper. Childhood heart disease in Australia: Current Practices and Future Needs. A report for HeartKids and Paediatric and Congenital Council of the Cardiac Society of Australia and New Zealand. Pennant Hills, NSW, Australia, HeartKids Australia.
- <sup>89</sup> Ibid.
- <sup>90</sup> KEMPNY, A., TUTAREL, O., DIMOPOULOS, K., UEBING, A., SHIINA, Y., ALONSO-GONZALEZ, R., LI, W., SWAN, L., BAUMGARTNER, H., GATZOULIS, M. & DILLER, G. (2016). "Meeting the challenge: The evolving global landscape of adult congenital heart disease." [International Journal of Cardiology](#) **168**: 5182-5189.
- <sup>91</sup> CORDINA, R., NASIR AHMAD, S., KOTCHETKOVA, I., EVEBORN, G., PRESSLEY, L., AYER, J., CHARD, R., TANOUS, D., ROBINSON, P., KILIAN, J., DEANFIELD, J. & CELERMAJER, D. (2018). "Management errors in adults with congenital heart disease: prevalence, sources, and consequences." [European Heart Journal](#) **39**: 982-989.
- <sup>92</sup> CSANZ (2013). Adult Congenital Heart Disease (ACHD) Recommendations for Standards of Care., The Cardiac Society of Australia and New Zealand.
- <sup>93</sup> NICOLAE, M., GENTLES, T., STRANGE, G., TANOUS, D., DISNEY, P., BULLOCK, A., GRIGG, L., CHEN, S.S.M., PRESSLEY, L., CORDINA, R., HORNUNG, T., O'DONNELL, C. & CELERMAJER, D.S. (in press). "Adult Congenital Heart Disease in Australia and New Zealand A Call for Optimal Care." [Heart, Lung and Circulation](#).
- <sup>94</sup> LEGGAT, S. (2011). The 2011 Childhood Heart Disease in Australia White Paper. Childhood heart disease in Australia: Current Practices and Future Needs. A report for HeartKids and Paediatric and Congenital Council of the Cardiac Society of Australia and New Zealand. Pennant Hills, NSW, Australia, HeartKids Australia.
- <sup>95</sup> Ibid.
- <sup>96</sup> NICOLAE, M. (2017). [Australian ACHD Centres Overview](#). ACHD Symposium 2017, Queenstown, New Zealand.
- <sup>97</sup> NICOLAE, M., GENTLES, T., STRANGE, G., TANOUS, D., DISNEY, P., BULLOCK, A., GRIGG, L., CHEN, S.S.M., PRESSLEY, L., CORDINA, R., HORNUNG, T., O'DONNELL, C. & CELERMAJER, D.S. (in press). "Adult Congenital Heart Disease in Australia and New Zealand A Call for Optimal Care." [Heart, Lung and Circulation](#).
- <sup>98</sup> MARELLI, A., THERRIEN, J., MACKIE, A., IONESCU-ITTU, R. & PILOTE, L. (2009). "Planning the specialized care of adult congenital heart disease patients: from numbers to guidelines; an epidemiologic approach." [American Heart Journal](#) **157**: 1-8.

---

<sup>99</sup> STOUT, K. K., DANIELS, C.J., ABOULHOSN, J.A., BOZKURT, B., BROBERG, C.S., COLMAN, J.M., CRUMB, S.R., DEARANI, J.A., FULLER, S., GURVITZ, M., KHAIRY, P., LANDZBERG, M.J., SAIDI, A., VALENTE, A.M. & VAN HARE, G.F. (2018). "2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease." Journal of the American College of Cardiology.

<sup>100</sup> NICOLAE, M., GENTLES, T., STRANGE, G., TANOUS, D., DISNEY, P., BULLOCK, A., GRIGG, L., CHEN, S.S.M., PRESSLEY, L., CORDINA, R., HORNUNG, T., O'DONNELL, C. & CELERMAJER, D.S. (in press). "Adult Congenital Heart Disease in Australia and New Zealand A Call for Optimal Care." Heart, Lung and Circulation.

<sup>101</sup> NICOLAE, M. (2017). Australian ACHD Centres Overview. ACHD Symposium 2017, Queenstown, New Zealand.

<sup>102</sup> AUSTRALIAN INSTITUTE OF HEALTH AND WELFARE (2018). Australia's health. Australia's health series. Canberra, AIHW AUSTRALIAN. **16**.

<sup>103</sup> CELERMAJER, D., NICOLAE, M., HORNUNG, T., O'DONNELL, C., JUSTO, R., BULLOCK, A., CHEUNG, M., COCHRANE, A., GENTLES, T., RAMSAY, J., SHOLLER, G. & WHEATON, G. (2013) Adult Congenital Heart Disease (ACHD) Recommendations for Standards of Care.

<sup>104</sup> NICOLAE, M., GENTLES, T., STRANGE, G., TANOUS, D., DISNEY, P., BULLOCK, A., GRIGG, L., CHEN, S.S.M., PRESSLEY, L., CORDINA, R., HORNUNG, T., O'DONNELL, C. & CELERMAJER, D.S. (in press). "Adult Congenital Heart Disease in Australia and New Zealand A Call for Optimal Care." Heart, Lung and Circulation.

<sup>105</sup> WARNES, C. A., WILLIAMNS, R.G., BASHORE, T.M., CHILD, J.S., CONNOLLY, H.M., DEARANI, J.A., DEL NIDO, P., FASULES, J.W., GRAHAM JR, T.P., HIIJAZI, Z.M., HUNT, S.A., KING, M.E., LANDZBERG, M.J., MINER, P.D., RADFORD, M.J., WALSH, E.P. & WEBB, G.D. (2008). "ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease: Executive Summary." Circulation **118**: 2395-2451.

<sup>106</sup> WILSON, M. and N. HUMBERSTONE (2016). "Adult Congenital Heart Disease Specifications." from [www.england.nhs.uk/wp-content/uploads/2018/08/Congenital-heart-disease-specification-adults.pdf](http://www.england.nhs.uk/wp-content/uploads/2018/08/Congenital-heart-disease-specification-adults.pdf).

<sup>107</sup> MARELLI, A., THERRIEN, J., MACKIE, A., IONESCU-ITTU, R. & PILOTE, L. (2009). "Planning the specialized care of adult congenital heart disease patients: from numbers to guidelines; an epidemiologic approach." American Heart Journal **157**: 1-8.

<sup>108</sup> LEGGAT, S. (2011). The 2011 Childhood Heart Disease in Australia White Paper. Childhood heart disease in Australia: Current Practices and Future Needs. A report for HeartKids and Paediatric and Congenital Council of the Cardiac Society of Australia and New Zealand. Pennant Hills, NSW, Australia, HeartKids Australia.

<sup>109</sup> MATHUR, S. and J. SUTTON (2017). "Personalized medicine could transform healthcare." Biomedical Reports. Retrieved June 2018, from <https://www.ncbi.nlm.nih.gov/pubmed/28685051>.

<sup>110</sup> LAWOKO, S. (2007). "Factors influencing satisfaction and well-being among parents of congenital heart disease children: development of a conceptual model based on the literature review." Scandinavian Journal of Caring Sciences **21**: 106-117.

<sup>111</sup> MITCHELL, A., BELTON, S., JOHNSTON, V., GONDARRA, W. & RALPH, A. (2018). "'The Heart Sickness:' Young Aboriginal People's Understanding of Rheumatic Fever." Medical Anthropology. from <https://www.tandfonline.com/action/showCitFormats?doi=10.1080%2F01459740.2018.1482549>.

<sup>112</sup> WARK, E., HODDER, Y., WOODS, C. & MAGUIRE, G. (2013). "Patient and health-care impact of a pilot rheumatic heart disease screening program." Journal of Paediatrics and Child Health **49**: 297-302.

- 
- <sup>113</sup> LAWOKO, S. (2007). "Factors influencing satisfaction and well-being among parents of congenital heart disease children: development of a conceptual model based on the literature review." Scandinavian Journal of Caring Sciences **21**: 106-117.
- <sup>114</sup> MITCHELL, A., BELTON, S., JOHNSTON, V., GONDARRA, W. & RALPH, A. (2018). "'The Heart Sickness:' Young Aboriginal People's Understanding of Rheumatic Fever." Medical Anthropology. from <https://www.tandfonline.com/action/showCitFormats?doi=10.1080%2F01459740.2018.1482549>.
- <sup>115</sup> WARK, E., HODDER, Y., WOODS, C. & MAGUIRE, G. (2013). "Patient and health-care impact of a pilot rheumatic heart disease screening program." Journal of Paediatrics and Child Health **49**: 297-302.
- <sup>116</sup> CALLUS, E. and G. PRAVETTONI (2018). "The Role of Clinical Psychology and Peer to Peer Support in the Management of Chronic Medical Conditions - A Practical Example With Adults With Congenital Heart Disease." Frontiers in Psychology **9**: 731.
- <sup>117</sup> HALL, S. L., RYAN, J., BEATTY, J. & GRUBBS, L. (2015) Recommendations for peer-to-peer support for NICU parents. Journal of Perinatology **35**, S19-S13 DOI: 10.1038/jp.2015.143.
- <sup>118</sup> CHONG, L., FITZGERALD, D., CRAIG, J., MANERA, K., HANSON, C., CELERMAJER, D., AYER, J., KASPARIAN, N. & TONG, A. (2017). "Children's experiences of congenital heart disease: a systematic review of qualitative studies." European Journal of Pediatrics **177**: 319-336.
- <sup>119</sup> GALKAL (2018). Project Unite – A national picture of conditions and charities. A report for HeartKids. Sydney, NSW, Australia, GALKAL Independent Australian Strategy and Research Company.
- <sup>120</sup> CHONG, L., FITZGERALD, D., CRAIG, J., MANERA, K., HANSON, C., CELERMAJER, D., AYER, J., KASPARIAN, N. & TONG, A. (2017). "Children's experiences of congenital heart disease: a systematic review of qualitative studies." European Journal of Pediatrics **177**: 319-336.
- <sup>121</sup> INGLES, J., SARINA, T., KASPARIAN, N. & SEMSARIAN, C. (2013). "Psychological wellbeing and posttraumatic stress associated with implantable cardioverter defibrillator therapy in young adults with genetic heart disease." International Journal of Cardiology **168**: 3779-3784.
- <sup>122</sup> KASPARIAN, N., WINLAW, D. & SHOLLER, G. (2016). "'Congenital heart health': how psychological care can make a difference." The Medical Journal of Australia **205**: 104-107.
- <sup>123</sup> KOVACS, A. H., SAIDI, A. S., KUHL, E. A., SEARS, S. F., SILVERSIDES, C., HARRISON, J. L., ONG, L., COLMAN, J., OECHSLIN, E. & NOLAN, R. P. (2009). "Depression and anxiety in adult congenital heart disease: predictors and prevalence." International Journal of Cardiology **137**: 158-164.
- <sup>124</sup> STEPTOE, A., MOHABIR, A., MAHON, N. & MCKENNA, W. (2000). "Health related quality of life and psychological wellbeing in patients with dilated cardiomyopathy." Heart **83**: 645-650.
- <sup>125</sup> WILSON, W., SMITH-PARRISH, M., MARINO, B. & KOVACS, A. (2015). "Neurodevelopmental and psychosocial outcomes across the congenital heart disease lifespan." Progress in Pediatric Cardiology **39**: 113-118.
- <sup>126</sup> KASPARIAN, N., WINLAW, D. & SHOLLER, G. (2016). "'Congenital heart health': how psychological care can make a difference." The Medical Journal of Australia **205**: 104-107.
- <sup>127</sup> WOOLF-KING, S. E., ANGER, A., ARNOLD, E. A., WEISS, S. J. & TEITEL, D. (2017). "Mental health among parents of children with critical congenital heart defects: a systematic review." Journal of the American Heart Association **6**(2): e004862.

---

<sup>128</sup> CHONG, L., FITZGERALD, D., CRAIG, J., MANERA, K., HANSON, C., CELERMAJER, D., AYER, J., KASPARIAN, N. & TONG, A. (2017). "Children's experiences of congenital heart disease: a systematic review of qualitative studies." European Journal of Pediatrics **177**: 319-336.

<sup>129</sup> INGLES, J., SARINA, T., KASPARIAN, N. & SEMSARIAN, C. (2013). "Psychological wellbeing and posttraumatic stress associated with implantable cardioverter defibrillator therapy in young adults with genetic heart disease." International Journal of Cardiology **168**: 3779-3784.

<sup>130</sup> KASPARIAN, N., WINLAW, D. & SHOLLER, G. (2016). "'Congenital heart health': how psychological care can make a difference." The Medical Journal of Australia **205**: 104-107.

<sup>131</sup> KOVACS, A. H., SAIDI, A. S., KUHL, E. A., SEARS, S. F., SILVERSIDES, C., HARRISON, J. L., ONG, L., COLMAN, J., OECHSLIN, E. & NOLAN, R. P. (2009). "Depression and anxiety in adult congenital heart disease: predictors and prevalence." International Journal of Cardiology **137**: 158-164.

<sup>132</sup> STEPTOE, A., MOHABIR, A., MAHON, N. & MCKENNA, W. (2000). "Health related quality of life and psychological wellbeing in patients with dilated cardiomyopathy." Heart **83**: 645-650.

<sup>133</sup> WILSON, W., SMITH-PARRISH, M., MARINO, B. & KOVACS, A. (2015). "Neurodevelopmental and psychosocial outcomes across the congenital heart disease lifespan." Progress in Pediatric Cardiology **39**: 113-118.

<sup>134</sup> DENNISS, D. L., SHOLLER, G.F., COSTA, D.S.J., WINLAW, D.S & KASPARIAN, N.A. (in press) Need for Routine Screening of Health-Related Quality of Life in Families of Young Children with Complex Congenital Heart Disease. Journal of Pediatrics DOI: <https://doi.org/10.1016/j.jpeds.2018.09.037>

<sup>135</sup> KASPARIAN, N., WINLAW, D. & SHOLLER, G. (2016). "'Congenital heart health': how psychological care can make a difference." The Medical Journal of Australia **205**: 104-107.

<sup>136</sup> Ibid.

<sup>137</sup> STATE OF QUEENSLAND (QUEENSLAND HEALTH) (2018). "Supporting the long-term developmental needs of children with congenital heart disease and their families." from <https://www.childrens.health.qld.gov.au/wp-content/uploads/PDF/pink-book.pdf>.

<sup>138</sup> MARINO, B., LIPKIN, P., NEWBURGER, J., PEACOCK, G., GERDES, M., GAYNOR, J., MUSSATTO, K., UZARK, K., GOLDBERG, C., JOHNSON WH JR, LI, J., SMITH, S., BELLINGER, D. & MAHLE, W. (2012). "Neurodevelopmental outcomes in children with congenital heart disease: evaluation and management: a scientific statement from the American Heart Association." Circulation **126**: 1143-1172.

<sup>139</sup> AUSTRALIAN INSTITUTE OF HEALTH AND WELFARE (2013). Rheumatic heart disease and acute rheumatic fever in Australia: 1996–2012. Cardiovascular disease series. Canberra, ACT, Australia, Australian Government. **Cat. no. CVD 60**.

<sup>140</sup> AUSTRALIAN INSTITUTE OF HEALTH AND WELFARE (2017). Aboriginal and Torres Strait Islander health performance framework 2017: supplementary online tables. Canberra, ACT, Australia, Australian Government. **Cat. no. WEB 170**.

<sup>141</sup> ARONEY, C., AYLWARD, P., CHEW, D., HUANG, N., KELLY, A., WHITE, H. & WILSON, M. (2008). "2007 addendum to the National Heart Foundation of Australia/Cardiac Society of Australia and New Zealand guidelines for the management of acute coronary syndromes 2006." Medical Journal of Australia **188**: 302-303.

- 
- <sup>142</sup> AUSTRALIAN INSTITUTE OF HEALTH AND WELFARE (2016). Better Cardiac Care measures for Aboriginal and Torres Strait Islander people: second national report 2016. Canberra, ACT, Australia, Australian Institute of Health and Welfare.
- <sup>143</sup> ILTON, M., WALSH, W., BROWN, A., TIDEMAN, P., ZELTZ, C. & WILSON, J. (2014). "A framework for overcoming disparities in management of acute coronary syndromes in the Australian Aboriginal and Torres Strait Islander population. A consensus statement from the National Heart Foundation of Australia." Medical Journal of Australia **200**: 639-643.
- <sup>144</sup> BROWN, A. and L. KRITHARIDES (2017). "Overcoming cardiovascular disease in Indigenous Australians." Medical Journal of Australia **206**: 10-12.
- <sup>145</sup> MITCHELL, A., BELTON, S., JOHNSTON, V., GONDARRA, W. & RALPH, A. (2018). "The Heart Sickness: Young Aboriginal People's Understanding of Rheumatic Fever." Medical Anthropology. from <https://www.tandfonline.com/action/showCitFormats?doi=10.1080%2F01459740.2018.1482549>.
- <sup>146</sup> RHDAUSTRALIA (2012). Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition). Quick reference guides: RHD Australia (ARF/RHD writing group). National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand.
- <sup>147</sup> GURVITZ, M. and A. SAIDI (2014). "Transition in congenital heart disease: it takes a village." Heart **100**: 1075-1076.
- <sup>148</sup> WILLIAMS, R. (2015). "Transitioning youth with congenital heart disease from pediatric to adult healthcare." Journal of Pediatrics **166**: 15-19.
- <sup>149</sup> AUSTRALIAN RESEARCH COUNCIL (2018). "ARC Medical Research Policy." Retrieved September 2018, from <https://www.arc.gov.au/policies-strategies/policy/arc-medical-research-policy/arc-medical-research-policy-version-20181>.
- <sup>150</sup> AUSTRALIAN RESEARCH COUNCIL (2015). "Research Impact Principles and Framework." Retrieved October 2018, from <https://www.arc.gov.au/policies-strategies/strategy/research-impact-principles-framework>.
- <sup>151</sup> NATIONAL HEALTH AND MEDICAL RESEARCH COUNCIL (2016). "Statement on Consumer and Community involvement in Health and Medical Research." from <https://nhmrc.gov.au/about-us/publications/statement-consumer-and-community-involvement-health-and-medical-research>.
- <sup>152</sup> WORLD HEALTH ORGANIZATION (2018). "Public health surveillance." Retrieved September 2018, from [https://www.who.int/topics/public\\_health\\_surveillance/en/](https://www.who.int/topics/public_health_surveillance/en/).
- <sup>153</sup> AUSTRALIAN RESEARCH COUNCIL (2018). "ARC Medical Research Policy." Retrieved September 2018, from <https://www.arc.gov.au/policies-strategies/policy/arc-medical-research-policy/arc-medical-research-policy-version-20181>.
- <sup>154</sup> AUSTRALIAN HEALTH MINISTERS' ADVISORY COUNCIL (2017). National Strategic Framework for Chronic Conditions. Canberra, ACT, Australia, Australian Government.
- <sup>155</sup> AUSTRALIAN COMMISSION ON SAFETY AND QUALITY IN HEALTH CARE (2018). "National arrangements for clinical quality registries." Retrieved October 2018, from <https://www.safetyandquality.gov.au/our-work/information-strategy/clinical-quality-registries/>
- <sup>156</sup> NATIONAL HEALTH AND MEDICAL RESEARCH COUNCIL (2015). "What is a clinical trial? ." Retrieved October 2018, from <https://www.australianclinicaltrials.gov.au/what-clinical-trial>.

---

<sup>157</sup> NATIONAL HEALTH AND MEDICAL RESEARCH COUNCIL (2016). "Statement on Consumer and Community involvement in Health and Medical Research." from <https://nhmrc.gov.au/about-us/publications/statement-consumer-and-community-involvement-health-and-medical-research>.

<sup>158</sup> WORLD HEALTH ORGANIZATION (2018). "WHO definitions of genetics and genomics." Retrieved October 2018, from <http://www.who.int/genomics/geneticsVSgenomics/en/>.

<sup>159</sup> Ibid.

<sup>160</sup> HEALTH ISSUES CENTRE (2018). "What are the types of healthcare? Australia's three-tiered healthcare system." Retrieved October 2018, from <https://healthissuescentre.org.au/consumers/health-care-in-australia/what-are-the-types-of-health-care>.

<sup>161</sup> AUSTRALIAN HEALTH MINISTERS' ADVISORY COUNCIL (2017). National Strategic Framework for Chronic Conditions. Canberra, ACT, Australia, Australian Government.

<sup>162</sup> AUSTRALIAN RESEARCH COUNCIL (2015). "Research Impact Principles and Framework." Retrieved October 2018, from <https://www.arc.gov.au/policies-strategies/strategy/research-impact-principles-framework>.

<sup>163</sup> AUSTRALIAN HEALTH MINISTERS' ADVISORY COUNCIL (2017). National Strategic Framework for Chronic Conditions. Canberra, ACT, Australia, Australian Government.

<sup>164</sup> HEALTH ISSUES CENTRE (2018). "What are the types of healthcare? Australia's three-tiered healthcare system." Retrieved October 2018, from <https://healthissuescentre.org.au/consumers/health-care-in-australia/what-are-the-types-of-health-care>.

<sup>165</sup> AUSTRALIAN COMMISSION ON SAFETY AND QUALITY IN HEALTHCARE (2013). "Clinical Care Standards Frequently asked questions (FAQs) for clinicians and health services." from <https://safetyandquality.gov.au/wp-content/uploads/2013/01/PDF-180KB.pdf>.

<sup>166</sup> WORLD HEALTH ORGANIZATION (2018). "Public health surveillance." Retrieved September 2018, from [https://www.who.int/topics/public\\_health\\_surveillance/en/](https://www.who.int/topics/public_health_surveillance/en/).

<sup>167</sup> HEALTH ISSUES CENTRE (2018). "What are the types of healthcare? Australia's three-tiered healthcare system." Retrieved October 2018, from <https://healthissuescentre.org.au/consumers/health-care-in-australia/what-are-the-types-of-health-care>.

<sup>168</sup> THE ROYAL CHILDREN'S HOSPITAL MELBOURNE (2018). "Transition Support Service - For young people." from [https://www.rch.org.au/transition/for\\_young\\_people](https://www.rch.org.au/transition/for_young_people).