

Beyond the Heart: Transforming Care

CHILDHOOD HEART DISEASE NATIONAL ACTION PLAN 2018

Draft for Public Consultation

September 2018

DRAFT

“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, Chairperson, HeartKids

“To the CHD 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

Development of the Childhood Heart Disease National Action Plan 2018 was led by HeartKids Ltd with funding from the Australian Government Department of Health.

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INTRODUCTION

Goal

People with childhood heart disease live longer, healthier and more productive lives through effective management of childhood heart disease across the life course.

Background – About this Action Plan

Beyond the Heart: Transforming Care – Childhood Heart Disease National Action Plan 2018 (the Action Plan) is the overarching plan to reduce the impact of childhood heart disease in Australia. The Plan is intended to minimise the long-term adverse medical, psychological and economic effects that may undermine the successes of early treatment and prevent individuals and families being their best and most productive selves throughout their life.

The Action Plan sets out the priority actions and outcomes to achieve the Goal that “People with childhood heart disease live longer, healthier and more productive lives through effective management of childhood heart disease across the life course.”

This Action Plan is the first nationally coordinated effort to tackle childhood heart disease.

The Action Plan draws on international progress, existing national hubs of excellence and builds on the momentum of recent initiatives; including the recently established Congenital Heart Disease Registry and the 2011 White Paper (Leggat, 2011). The White Paper provided a review of the delivery of healthcare in Australia to children with childhood heart disease – acquired and congenital heart disease. It found there were significant areas of deficit within service delivery and family support and provided a number of recommendations for planning improved healthcare delivery and support. Now almost a decade later many of these issues retain currency and are under intense international focus, and action to redress under resourcing of childhood heart disease care is critical.

The Action Plan is a call to action to:

- Enable individuals and families affected by childhood heart disease to be their best and most productive selves throughout their life
- Look beyond the heart and transform care so that it is life-long, holistic and person and family-centred
- Initiate and accelerate research breakthroughs to change lives
- Secure best practice now and in the future.

Many individuals and organisations contributed time and expertise to the development of the plan: including people with childhood heart disease; parents, carers and bereaved families; health professionals; key medical and cardiac organisations; the research community and the Australian Government and State and Territory Governments.

Beyond the Heart: Transforming Care - Childhood Heart Disease National Action Plan 2018

A considered and compassionate response to the major health burden of childhood heart disease is outlined, with recommendations for investment in three broad areas: Management, Care and Support; Supportive Communities; and Research. The plan addresses childhood heart disease encompassing congenital and acquired heart disease throughout, with the exception of Research that relates specifically to congenital heart disease.

The Action Plan aligns with and supports the policy directions in the National Strategic Framework for Chronic Conditions (the Framework) (Australian Health Ministers' Advisory Council, 2017), which provides the overarching national policy for the prevention and management of chronic conditions in Australia. The Action Plan also aligns and supports key national health strategies including the renewed commitment to Closing the Gap, and the recently announced Rheumatic Heart Disease (RHD) Strategy.

What is Childhood heart disease?

For the purposes of this Action Plan, **childhood heart disease** (CHD) is a general term for a range of conditions that affect the normal workings of the heart. Childhood heart disease can be present at birth (congenital) or acquired during childhood. Childhood heart disease is a chronic condition that requires complex, specialised care across the life course.

Childhood heart disease 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 childhood heart disease is unrelated to lifestyle.

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).

Congenital heart disease is any defect of the heart or major blood vessels that is present at birth. Some defects are mild and cause no significant disturbance to the way the heart functions. However, more than half of all children with congenital heart disease will require treatment at some stage of life.

An **acquired heart defect** occurs when a person's heart is normal at birth but later develops a problem. Illnesses that can lead to a heart problem include myocarditis (inflammation of the heart muscle), cardiomyopathy (disease of the heart muscle), rheumatic heart disease (a disease of the heart valves that can follow Strep A bacterial infection) and Kawasaki disease (a disease with fever, rash and swollen lymph glands that may affect the heart).

Most cases of congenital heart disease have no known cause.

Conversely, rheumatic heart disease, an acquired heart defect which occurs in Australia almost exclusively in Aboriginal and Torres Strait Islander communities, is entirely preventable.

The causes of congenital heart disease remain largely unclear. Some of the known causes include genes, environmental factors and other factors relating to maternal health. It is estimated that 20% of cases have a genetic cause. In around eight out of 10 cases, the reason is unknown (Better Health Channel, 2014). There is no evidence for effective interventions to prevent congenital heart disease.

Conversely, rheumatic heart disease, a preventable acquired heart defect, is caused by an abnormal immune reaction to Strep 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. In Australian children rheumatic heart disease occurs almost exclusively in Aboriginal and Torres Strait Islander communities and is entirely preventable.

Surgery can reduce the impact of the disease although it doesn't provide a 'cure'. People with childhood heart disease have complex, life-long needs.

Treatment is available for most congenital heart defects which can reduce the impact of the disease, however there is no cure. More than half of all children with a heart defect have a condition that is serious enough to require treatment (Leggat, 2011). Evidence is emerging that childhood surgical repair of the heart has longer-term impacts and outcomes. However, early diagnosis and treatment can lead to better long-term management and health outcomes, including quality of life.

AUSTRALIAN SITUATION

The Changing Landscape of Childhood Heart Disease

Thanks to medical advances in Australia and internationally over the past 75 years, childhood heart disease survival rates have improved dramatically and many childhood heart problems that were fatal are now chronic medical conditions requiring ongoing management. The decline in mortality has shifted dramatically over the past two decades, and the majority (>95%) of people with childhood heart disease now survive into adulthood (Le Gloan et al., 2011, Khairy et al., 2010).

The gains in survival for childhood heart disease are a triumph, but paradoxically, they bring new challenges. Earlier diagnosis, more complex treatment choices, longer survivorship and a need for transition from paediatric to adult cardiac services lead us into new territory and pose new threats and demands on Australia's health care system, educational institutions, workforce and broader economy.

Childhood heart disease places a significant burden on people affected by the disease, Australia's health care system, educational institutions, workforce and broader economy.

There remains a focus on childhood, where the intensity of interventions occur – approximately half of all children with congenital heart disease and a significant lesion have surgery/catheter intervention within their first year of life (Sholler et al., 2011). Yet the surgical successes of childhood heart disease, which has led to longer survivorship, means that we must now look beyond childhood and consider whole of life care, from diagnosis, infancy, childhood, adolescence, and adulthood.

Currently in Australia, approximately 65,000 - 90,000 children and adults live with childhood heart disease. A lack of definitive research on the burden of disease suggest the number could be far higher and is predicted to increase. For the first time in history, adults living with childhood heart disease outnumber children with the disease. This large and growing population require lifelong and highly specialised medical care. This changing demographic and epidemiology of childhood heart disease has shifted a large portion of the burden of disease into the adult population, with little understanding of the cost to patients, the health system, education system, workforce, and national economy.

Longer survivorship means that we must now look beyond childhood and consider whole of life care, from diagnosis, infancy, childhood, adolescence, and adulthood.

Surgery can reduce the impact of the disease although it doesn't provide a 'cure', and there are a distinct range of issues in adult patients after surgery, including mental health problems and neurodevelopmental impairment. Care for childhood heart disease must look beyond the heart and take a more holistic approach that improves physical health alongside

mental, cognitive, social and emotional health. Beyond care, we must also look at how people with childhood heart disease and their families can lead active, fulfilling lives and most productively contribute to the Australian economy.

Treatment doesn't provide a 'cure' and a distinct range of issues in adult patients exist after surgery; a holistic approach that improves physical health alongside mental, cognitive and emotional health is required.

Evidence is growing that childhood heart disease not only impacts the quality of life of individuals but has a significant impact on their families including parents, carers and siblings. It is critical that Australia's health system is shaped to recognise and value the needs of individuals, their carers and their families, and that person-centred and family-centred approaches underpin childhood heart disease care.

Person-centred and family-centred approaches must underpin childhood heart disease care.

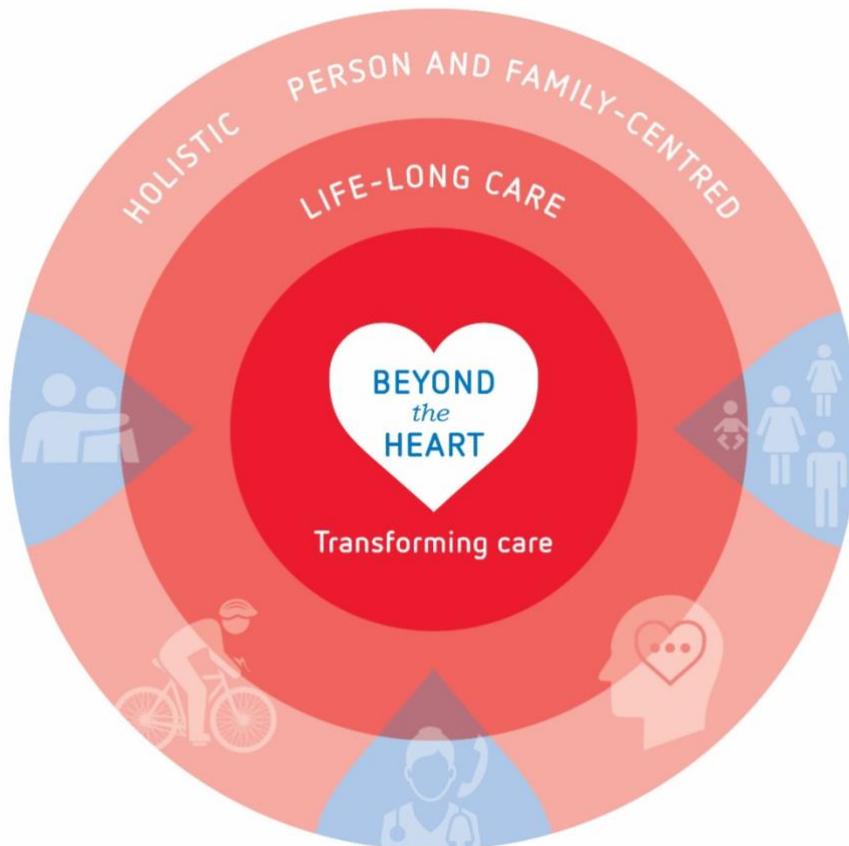
Despite excellence in some areas of cardiac care, there is an urgent need for improved healthcare delivery and support. The growing number of people affected by childhood heart disease and the rapid growth in the number of adults living with childhood heart disease is placing pressure on Australia's health care system and highlighting gaps in service delivery and family support. Additionally, due to a lack of coordinated national effort to date, childhood heart disease is characterised a lack of data, the absence of national standards of care, and deficiencies in specialist adult CHD services and workforce shortages, which are limiting patient access and comprising quality of care.

Despite excellence in some areas of cardiac care in Australia, there are significant gaps in the continuity and completeness of service delivery that require urgent action.

Beyond the Heart: Transforming Care – Childhood Heart Disease National Action Plan 2018 provides a blueprint for Australia to transform care and secure the best possible outcomes for people affected by childhood heart disease, now and in the future.

This Action Plan responds to the changing landscape of childhood heart disease. It delivers on the field's strong and united call for Australia to transform childhood heart disease care and provides a contemporary focus on providing whole of life care that is holistic, person- and family-centred to enable people with childhood heart disease to lead fuller, active and productive lives (Figure 1).

Figure 1: Australia's contemporary approach to tackling childhood heart disease



Why Childhood Heart Disease matters

Childhood heart disease has a major impact on individuals and their families, the health system and the broader community. People with childhood heart disease require lifelong and highly specialised medical care and support, and many are exposed to significant hardship and suffering.

Key statistics about the health, social and economic impact of childhood heart disease are presented in Box 1.

Further detailed information is provided in Appendix 1: *The size of the problem*.

Box 1: Summary of the health, social and economic impact of Childhood Heart Disease

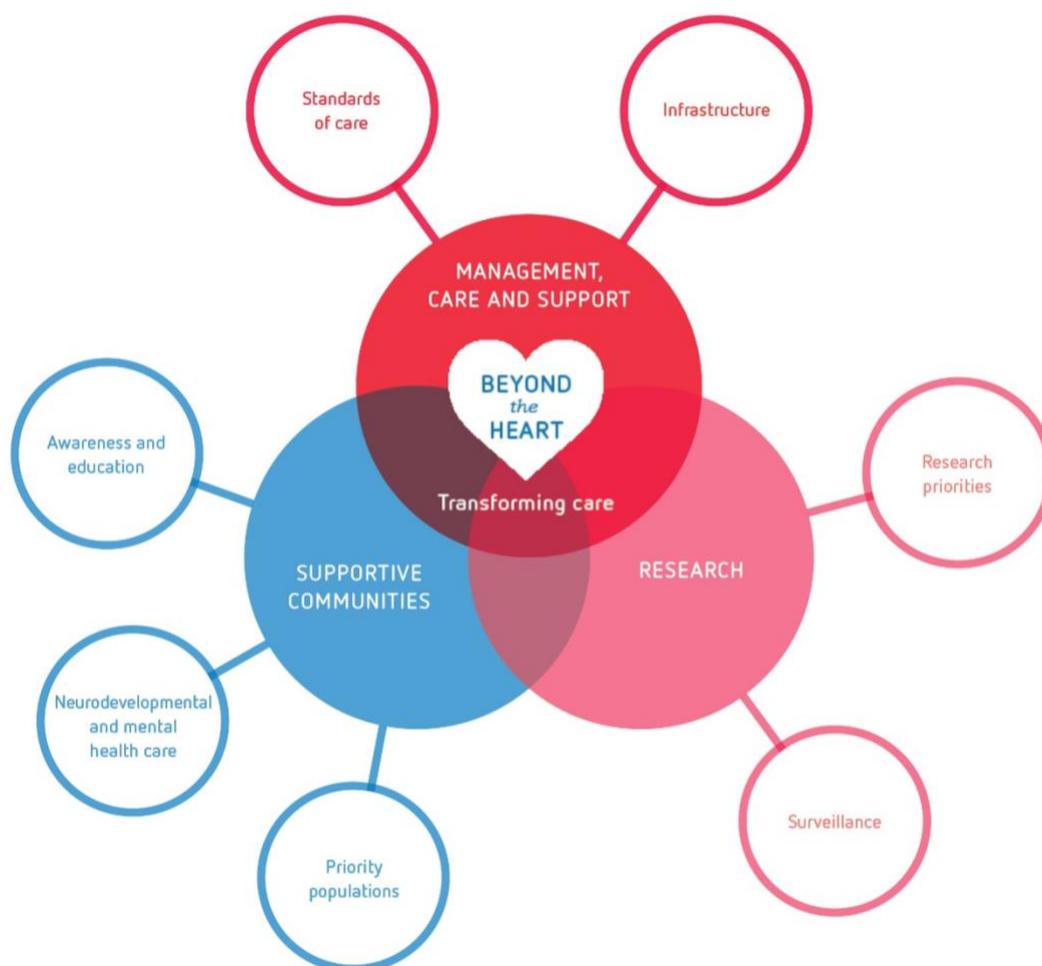
- Around 65,000 - 90,000 Australian children and adults live with childhood heart disease
- Congenital heart defects are the most common types of birth defects; eight babies are born with a congenital heart condition every day
- Congenital heart disease is one of the leading causes of death and hospitalisation in infants
- Four young lives a week are lost every week
- Aboriginal and Torres Strait Islander people experience higher rates of congenital and acquired heart disease and worse outcomes compared with non-Indigenous Australians.
- People with childhood heart disease 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)
- People with childhood heart disease are at a greater risk of neurodevelopmental impairment and disability including developmental delay and other learning difficulties
- Childhood heart disease poses a substantial economic burden on our health care system and results in an enormous financial burden for the majority of families at a very vulnerable time.

THE APPROACH

This Action Plan articulates a Goal supported by three high-level priorities that together will transform the care of people affected by childhood heart disease: Management, Care and Support; Supportive Communities; and Research (Figure 2). Each priority has a number of related and interdependent focus areas. The focus areas have been informed by evidence and the expert advice of the Childhood Heart Disease National Action Plan Steering Committee and Expert Working Groups and consultations with people affected by childhood heart disease and other stakeholders.

This Action Plan includes principles to guide action and enablers that will strengthen efforts across each of the priorities.

Figure 2: Childhood Heart Disease National Action Plan 2018



Goal

People with childhood heart disease live longer, healthier and more productive lives through effective management of childhood heart disease across the life course.

Summary Table of Recommended Actions

	Focus Area	Recommended Action
Management, Care and Support	1. Standards of Care	1.1 Establish a Childhood Heart Disease Taskforce to oversee the development of standards of care for childhood heart disease
		1.2 Develop national Standards of Care for Childhood Heart Disease
		1.3 Develop authoritative national clinical practice guidelines on childhood heart disease for health services and health practitioners throughout Australia
	2. Infrastructure	2.1 Develop a Childhood Heart Disease Infrastructure Plan
		2.2 Invest in technological infrastructure
		2.3 Deliver education, training and support for health professionals
		2.4 Support the management of childhood heart disease in primary care
		2.5 Disseminate best practice information and approaches throughout services in Australia
	Supportive communities	3. Awareness and Education
3.2 Increase access to ongoing support, by funding inpatient and outpatient Community (Peer) Support team members		
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 health care		4.1 Develop Standards of Mental Health Care for Childhood Heart Disease
		4.2 Develop Standards of Neurodevelopmental Care for Childhood Heart Disease
		4.3 Establish clear models of care
5. Priority populations		5.1 Fund CHD Cardiac Coordinators in each jurisdiction
		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

		5.5 Investigate options for reducing barriers to accessing care and supporting CHD families
Research	6. Research priorities	6.1 Longitudinal impact and informed practice with the Bi-National Congenital Heart Disease Registry
		6.2 Preventing Neurodevelopmental and Mental Health Complications
		6.3 Understanding CHD genetics to provide personalised medicine and support for those impacted by CHD - National Neonatal CHD Cohort Study
		6.4 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
		7.2 Continued funding of the National CHD Survey
		7.3 Develop an annual report card to monitor progress against this Action Plan

Principles

In line with the Framework (Australian Health Ministers' Advisory Council, 2017), this Action Plan is underpinned by eight guiding principles. These principles are expected to guide future action to address childhood heart disease and other chronic conditions.

1. Equity – all Australians, including Aboriginal and Torres Strait Islander people and other priority populations, receive culturally safe and appropriate, high quality health care.
2. Collaboration and partnerships – identify linkages and act upon opportunities to cooperate and partner responsibly to achieve greater impacts than can occur in isolation.
3. Access – high standard, appropriate support and services are available, accessible and affordable for all Australians.
4. Evidence-based – rigorous, relevant and current evidence informs best practice and strengthens the knowledge base to effectively prevent and manage chronic conditions.
5. Person-centred approaches – the health system is shaped to recognise and value the needs of individuals, their carers and their families, to provide holistic care and support.
6. Sustainability – strategic planning and responsible management of resources delivers long-term improved health outcomes.
7. Accountability and transparency – decisions and responsibilities are clear and accountable and achieve best value with public resources.

8. Shared responsibility – all parties understand, accept and fulfil their roles and responsibilities to ensure enhanced health outcomes for all Australians (Australian Health Ministers' Advisory Council, 2017).

Enablers

In line with the Framework (Australian Health Ministers' Advisory Council, 2017), this Action Plan draws on seven enablers that will assist in achieving the Goal of this plan:

- Governance and leadership – supports evidence-based shared decision-making and encourages collaboration to enhance health system performance.
- Health workforce – a suitably trained, resourced and distributed workforce is supported to work to its full scope of practice and is responsive to change.
- Health literacy – people are supported to understand information about health and health care, to apply that information to their lives and to use it to make decisions and take actions relating to their health.
- Research – quality health research accompanied by the translation of research into practice and knowledge exchange strengthens the evidence base and improves health outcomes.
- Data and information – the use of consistent, quality data and real-time data sharing enables monitoring and quality improvement to achieve better health outcomes.
- Technology – supports more effective and accessible prevention and management strategies and offers avenues for new and improved technologically driven initiatives.

PRIORITY 1: MANAGEMENT, CARE AND SUPPORT

FOCUS AREA 1: STANDARDS OF CARE

For the purposes of this Action Plan,

Standards of Care are a national agreement of key 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 health care provision and provide a framework to ensure people receive the best possible care and outcomes. Standards define the type of care that a person with childhood heart disease can expect. They provide a guide to the organisation and delivery of care and the related service and resource requirements, including personnel and equipment that are needed for safety and excellence.

The standards of care in hospital and community settings are an important factor in the provision of seamless congenital cardiac pathways.

Development of Standards of Care for Childhood Heart Disease is a priority of this Action Plan, along with the establishment of a governing body – the Childhood Heart Disease Taskforce – to oversee the development and ongoing review of the Standards.

Australia has never had nationally agreed Standards of Care for childhood heart disease, which has disadvantaged not only people with childhood heart disease and their families, but also our health care system, educational institutions, workforce and broader economy.

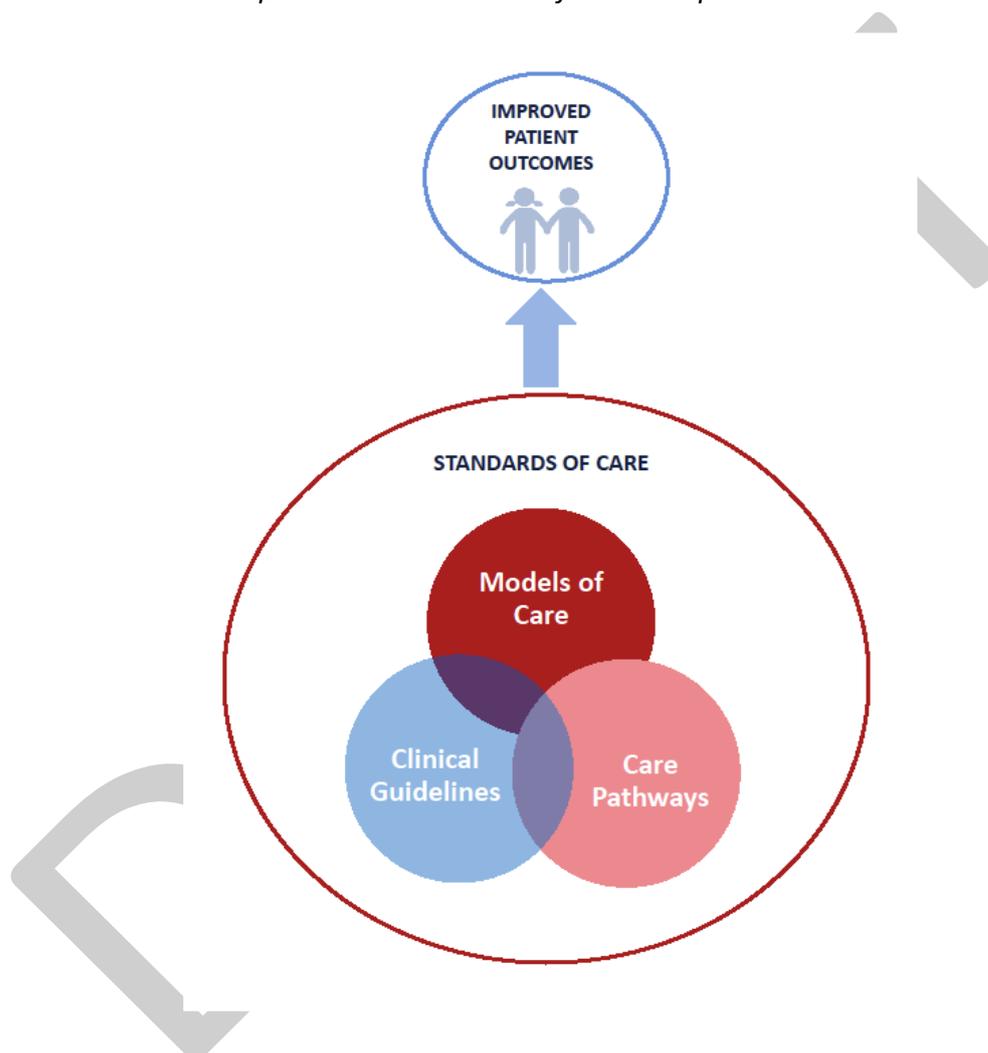
Despite excellence in many areas of cardiac care in Australia, there are a number of known and evolving points of vulnerability that compromise patient outcomes. The current care delivery process is fragmented at times and focussed on ‘moments of care’ rather than lifelong ‘continuity of care’. Securing quality care across the life course can be challenging and certain stages require urgent attention, such as transition of care from paediatric to adult cardiac health services, where over 50% of Australians with childhood heart disease are being ‘lost’ to care.

The integration and sustainability of services required to secure best practice now and, in the future, requires urgent action. The development of Childhood Heart Disease Standards of Care will ensure that all people with childhood heart disease have access to consistent, quality care across the life course.

Standards influence every aspect of care and represent a key step towards improving outcomes for people with childhood heart disease in Australia.

Strong quality standards will help Australia to sustain the best outcomes for all patients and reduce the physical, psychosocial, social and economic impact of childhood heart disease on individuals, families, the health system and the broader community. Figure 3 illustrates the relationship between Standards of Care and patient outcomes.

Figure 3: The relationship between Standards of Care and patient outcomes



A number of best practice examples already exist internationally, such as the NHS England Congenital Heart Disease Standards and Specifications (NHS England, 2016) and the Specialist Congenital Heart Disease standards for children and adults in Scotland (Scotland, 2018). Box 1 provides an overview of the categories contained in the Specialist Congenital Heart Disease standards for children and adults in Scotland.

Box 1: Categories of the Scottish Standards of Care

The Scottish standards encompass the whole patient pathway and are subdivided into categories:

- A – The Service Approach
- B – Staffing and Skills
- C – Facilities
- D – Interdependencies
- E – Training and Education
- F – Organisation, Governance and Audit
- G – Research
- H – Communication with Patients
- I – Transition
- J – Pregnancy and Contraception
- K – Fetal Diagnosis (paediatrics only)
- L – Palliative Care and Bereavement
- M – Dental (Scotland, 2018).

Standards of Care for Childhood Heart Disease will be tailored to the unique Australian context and will enhance existing cardiac services provided across the country. The standards will include components that build on existing international and Australian resources including care of adults with congenital heart disease [the Adult Congenital Heart Disease Recommendations for Standards of Care developed by the Cardiac Society of Australia and New Zealand (CSANZ, 2013)] and care for priority populations through the service standards for equitable cardiovascular care for Aboriginal and Torres Strait Islander People service standards (Brown et al., 2015).

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

Standards of Care intersects with each of the seven areas of focus of this Action Plan, particularly Focus Area 2: Infrastructure. Standards and specifications will inform future planning around infrastructure, including resource allocation. This includes addressing Australia's significant limitations in care and support capabilities due to workforce shortages and a lack of infrastructure and services.

“Standards of Care will secure the lifelong wellbeing of patients with childhood heart disease by building a defined base of excellence into the Australian Health System.”

Associate Professor Gary Sholler, Paediatric & Fetal Cardiologist, Chair, Standards of Care Expert Working Group

What will be different?

- People with childhood heart disease receive care of consistently high quality that is sustainable for the future
- Improved health outcomes for people with childhood heart disease and their families, including quality of life, physical health, mental health and wellbeing, and neurodevelopmental outcomes
- Lifelong opportunities for excellence in care, resulting in a reduced burden of childhood heart disease on the health care system, educational institutions, workforce and the broader economy
- Continued and embedded funding for the Childhood Heart Disease Taskforce.

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Recommended actions: Standards of Care

Action	Detail	Lead
1.1 Establish a Childhood Heart Disease Taskforce to oversee the development of standards of care for childhood heart disease	<p>The Taskforce will be established to oversee the development of Standards of Care for childhood heart disease. The Taskforce to be responsible for ongoing surveillance, review and periodic updates of the Standards.</p> <p>The Taskforce to be resourced with capacity to respond to areas of need identified during the development of the Standards, and to address gaps between the care defined by the Standards and current practice (e.g. development of models of care).</p> <p>The Taskforce to comprise a working group of clinicians, researchers, policy makers and consumers, including representation from Federal, State and Territory governments.</p>	Federal Government to establish Taskforce in consultation with experts in the field
1.2 Develop national Standards of Care for Childhood Heart Disease	<p>Develop a set of standards and service specifications that cover the entire patient pathway, from prevention, diagnosis, through to treatment, and on into care at home and end of life care, to make sure that every child, young person and adult with childhood heart disease, in every part of the country, will receive the same high standard of care.</p> <p>Standards of Care to consider Australia's unique geographical challenges and the needs of priority populations, and include:</p> <ul style="list-style-type: none"> • Organisation of care to meet current and future population needs • Service model • Care pathways to map the patient journey across primary, secondary and tertiary levels of care and community settings • Service specification standards and resource requirements, including personnel and equipment 	Childhood Heart Disease Taskforce

	<ul style="list-style-type: none"> • Resources and frameworks for minimum effective communication with patients, families, and other care providers, including priority populations • Information reporting requirements including patient satisfaction measure and clinical outcomes measures • Training, education and certification requirements for those involved in providing care. <p>Standards to include neurodevelopmental and mental health care standards and specifications (see Focus Area 4 in this Action Plan).</p>	
<p>1.3 Develop authoritative national clinical practice guidelines on childhood heart disease for health services and health practitioners throughout Australia</p>	<p>Following the development of Standards of Care, the Childhood Heart Disease Taskforce will work with experts to devise a road map and priorities for the development of national clinical practice guidelines and deliver a limited number of early versions of priority guidelines.</p> <p>Clinical Practice Guidelines will be developed over subsequent years, using the best evidence possible, for 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 integrative care for all stages of the care pathway and deliver quality and consistency for current and future generations.</p> <p>The development of National Health and Medical Research Council (NHMRC)-endorsed Clinical Practice Guidelines should include a review of existing 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 childhood heart disease for consideration in the Australian context. The development of guidelines for echocardiographic screening for RHD is one priority area for consideration.</p>	<p>Childhood Heart Disease Taskforce, in partnership with CSANZ and key stakeholders</p>

	Guidelines to be available in multiple formats including web-based, to maximise accessibility and uptake by the multidisciplinary care team.	
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FOCUS AREA 2: INFRASTRUCTURE

For the purposes of this Action Plan,

Infrastructure refers to the service and resource requirements needed to deliver quality care for people with childhood heart disease. Infrastructure includes personnel – the multidisciplinary workforce providing care, and the facilities, services and equipment that are needed for safety and excellence in childhood heart disease care.

Childhood heart disease infrastructure in Australia is under-resourced, both in terms of trained personnel and services, dedicated facilities and family support. Current deficits are expected to increase over time due to the growing population of adults with childhood heart disease that will place additional demands on the health care system.

Childhood heart disease infrastructure in Australia is under-resourced and inconsistent with international guidelines; there are significant workforce shortages and gaps in the continuity and completeness of service delivery.

Significant infrastructure deficiencies and inconsistencies with international guidelines were identified in the 2011 Childhood Heart Disease in Australia White Paper (Leggat, 2011), which reviewed the delivery of healthcare in Australia to people with childhood heart disease. The White Paper highlighted the substantial impact of workforce shortages and lack of services, including poorer health outcomes for Australians with childhood heart disease, and a growing burden on our health care system, educational institutions, workforce and broader economy.

The lack of adequate infrastructure has a catastrophic impact at certain stages of the care journey, such as transition of care from paediatric to adult cardiac health services where over 50% of Australians with childhood heart disease are being 'lost' to care. Importantly, the large and growing number of adults with childhood heart disease will result in an increase in demand for services and exacerbate infrastructure challenges. The lack of 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.

Now, almost a decade later few recommendations from the White Paper have been enacted. Action to redress the chronic under-resourcing of childhood heart disease care is critical.

“Paediatric cardiology services remain the mainstay of childhood heart disease management, but if adult services are inadequately planned and funded the benefits of

the large financial, medical and social investment made during childhood will be lost to avoidable mortality and morbidity in adolescence and adulthood.”

(Nicolae et al., (in press))

A review of State and Territory cardiovascular network plans and documents (where made available) reveals very little evidence of a planned approach to the infrastructure needs and requirements relating directly to childhood heart disease.

The delivery of care for people with childhood heart disease is enabled by existing infrastructure (e.g. TeleHealth, MyHealth record). The need for additional infrastructure relating directly to childhood heart disease will be informed by the new Standards of Care. The Standards and specifications provide a guide to the organisation and delivery of care and the related service and resource requirements, including personnel and equipment.

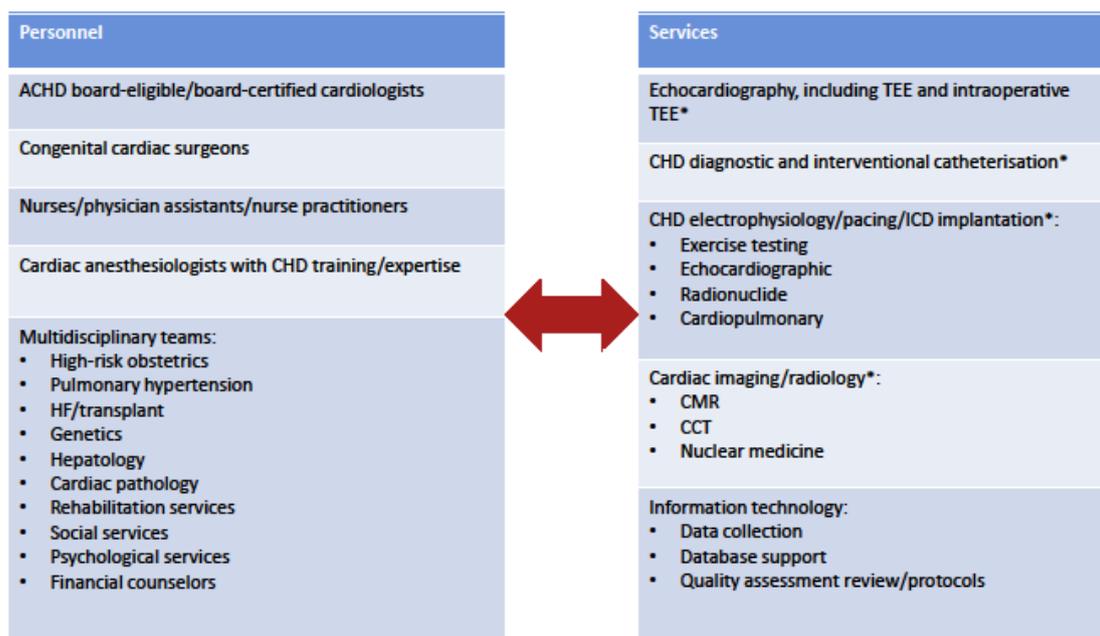
The new Standards of Care for Childhood Heart Disease will inform infrastructure planning and provide guidance for investment and action by State and Territory governments.

The national Standards of Care for Childhood Heart Disease (Focus Area 1 of this Action Plan) will form the basis of infrastructure planning relating directly to childhood heart disease, and feed into the development of an infrastructure plan. The infrastructure plan will redress Australia’s significant limitations in care and support capabilities and provide guidance for investment and action by State and Territory governments. A number of additional actions relating to childhood heart disease infrastructure are outlined below. Focus Area 5: Priority Populations 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.

Workforce

People with childhood heart disease have complex health needs and require lifelong care from a team of specialists in a range of institutions, both public and private. Figure 4 illustrates the infrastructure needs for Adult Congenital Heart Disease (ACHD) centres including key personnel, although this is limited to one tertiary setting. A much broader multidisciplinary group of professionals are involved in childhood heart disease care across the life course.

Figure 4: Key Personnel and Services Recommended for ACHD Programs (Stout et al., 2018)



*The modalities must be supervised/performed and interpreted by clinicians with expertise and/or training in CHD.

ACHD indicates adult congenital heart disease; CCT, cardiac computed tomography; CHD, congenital heart disease; CMR, cardiovascular magnetic resonance; HF, heart failure; ICD, implantable cardioverter-defibrillator; and TEE, transesophageal echocardiography.

Australia's current workforce is under-resourced to meet both current and future needs. Workforce shortages and skill gaps are limiting access and comprising patient quality of care (Kempny et al., 2016).

A critical issue is the shortage of adult congenital heart disease (ACHD) cardiologists. In 2010 there were approximately 30 paediatric cardiologists in Australia (Leggat, 2011), yet only 6 FTE cardiologists who care for adults with childhood heart disease (Nicolae, 2017, Nicolae et al., (in press)). This equates to 1 FTE ACHD specialist per 3.5 million population in Australia, in contrast to international recommendations of 1 FTE ACHD specialist per 1 million population or 1 ACHD centre per 2 million population (Stout et al., 2018, Marelli et al., 2009).

The recent NHS UK Review of the ACHD Services recommend at least 4 FTE congenital cardiologists and at least 4 FTE congenital surgeons for a Level 1 ACHD Specialist Centre (this is similar to a Comprehensive Adult Congenital Heart Centres (CACH) in Australia) (Wilson and Humberstone, 2016). Specific recommendations are also for Level 2 and 3 ACHD Centres (these are equivalent to the Regional Adult Congenital Heart Centres (RACH) in Australia).

In most CHD centres in Australia, there are currently fewer cardiologists, surgeons and other specifically trained health care professionals than recommended by international standards (Leggat, 2011). As a result, many adults with childhood heart disease have been lost to follow-up or are being cared for by general cardiologists with inadequate training resulting in a high risk for adverse clinical outcomes (Cordina et al., 2018).

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 and transition coordinators. Care coordinators that can work in the multidisciplinary team and coordinate and case manage people with childhood heart disease, particularly Aboriginal and Torres Strait Islander people and those living in regional, rural and remote areas, are particularly important and represent a large gap in Australia's current workforce.

No dedicated ACHD Centres in Australia have an attached psychologist or physiotherapist, and the majority have no Social Worker or even a Nurse Educator (Nicolae, 2017).

Additional workforce issues relate to the low level of childhood heart disease awareness, knowledge and capacity in primary care services, the lack of training pathways and training positions for ACHD cardiology, and a lack of training, education and certification requirements for those involved in providing care.

As part of the Infrastructure Plan, a workforce plan and further investment in workforce development are recommended to ensure Australia has an adequately trained, resourced and sustainable workforce to provide quality care for people with childhood heart disease.

Facilities, services and equipment

Childhood heart disease services in Australia are severely under-resourced, particularly in terms of dedicated facilities and family support.

Consistent with international best practice, a 'hub and spoke' model of care is recommended for congenital heart disease (Celermajer et al., 2013, Wilson and Humberstone, 2016, Warnes et al., 2001, Nicolae et al., (in press)), comprising centralised comprehensive centres (CACH) to provide care for people with moderate to severe spectrum of disease, and regional centres (RACH) for the milder spectrum of disease.

A critical issue is the shortage of dedicated facilities for childhood heart disease. There are currently only 5 designated CACH across Australia, although based on recommendations of having 1 Unit for every 2,000,000 (Marelli et al., 2009), Australia should have at least 8 CACH centres. There are currently no dedicated Regional ACHD Centres in Australia which puts increased pressure on the dedicated CACH centres, but also on the ACHD patients and their families who have to travel long distances.

Major issues for families include the cost of travel and accommodation when accessing care away from home, insufficient family facilities in hospitals, lack of access to respite care, and surgery cancellations as a consequence of insufficient resources in Paediatric Intensive Care Unit (PICU) and Intensive Care Unit (ICU) (Leggat, 2011). This is particularly challenging for people living in regional, rural and remote areas due to Australia's vast geography. Greater co-location of family support services within hospital settings would help to redress this.

No evidence can be found of the adaptation or uptake of e-health technologies to reduce the needs of rural and remote patients to travel (i.e. video conference).

There appears to be little forward planning or future proofing to address the emergence of new treatment options i.e. personalised medicine and the associated infrastructure costs over the next decade, as this becomes the norm.

What will be different?

- The health workforce is suitably trained, resourced and distributed to meet identified need
- The health workforce is working to its full scope of practice
- Effective multidisciplinary teams and health care partnerships in place
- Service delivery is coordinated and complete
- 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 childhood heart disease between Aboriginal and Torres Strait Islander people and non-Indigenous Australians is reduced.
- The disparity in health outcomes due to childhood heart disease between people living in regional, rural and remote areas and metropolitan areas is reduced.

Recommended actions: Infrastructure

Action	Detail	Lead
2.1 Develop a Childhood Heart Disease Infrastructure Plan	<p>Develop a national Childhood Heart Disease Infrastructure Plan to ensure Australia is able to provide quality and accessible care to meet current and future population needs.</p> <p>As part of the plan:</p> <ul style="list-style-type: none"> • Define a multidisciplinary care team for both paediatric and adult care settings • 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 • Review existing training (Vocational, Undergraduate and Post-Graduate, and Fellowship) to identify how future education, training and certification can improve health outcomes and meet the evolving needs of people with childhood heart disease. Opportunities include funded Fellowships; training in congenital cardiac surgery; and education, training and certification requirements for those involved in providing care. <p>The national Standards of Care for Childhood Heart Disease (Focus Area 1 of this Action Plan) will form the basis of infrastructure planning and provide guidance for investment and action by State and Territory governments.</p>	Childhood Heart Disease Taskforce, in partnership with key stakeholders
2.2 Invest in technological infrastructure	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 MyHealth Record.	Congenital Heart Alliance of Australia and New Zealand (CHAANZ)

	<p>Develop digital platforms to adequately capture outcomes.</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>Deliver education, training and support for health professionals to develop the workforce and its capacity to provide consistent, quality care for people with childhood heart disease.</p> <p>Education, training and support to focus on:</p> <ul style="list-style-type: none"> • Patient and family-centered care for people with childhood heart disease • The physical, psychological, social and cognitive impacts of childhood heart disease • Comorbidities, including neurological impairment • Culturally safe and appropriate communication and counselling techniques • Support for health professionals to minimise burnout and compassion fatigue • Referral pathways • Improving knowledge of primary care doctors and general cardiologists about the special needs of this group <p>Deliver education programs to support primary care, rural health professionals and community based allied health staff, and enhance the needs of workers that support priority populations (Focus Area 5 of this Action Plan) including the development of culturally safe and accessible education for Aboriginal and Torres Strait Islander Health Workers.</p> <p>Develop resources and tools to support health professionals 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>HeartKids in conjunction with relevant peak bodies, such as the Royal Australian College of General Practitioners' Continuing Professional Development Program and RHD Australia</p>

	<p>Education, training and support to engage the broader cardiology community and cardiologists working in both the public and private system.</p> <p>Education, training and support to align with Standards of Care for Childhood Heart Disease.</p>	
2.4 Support the management of childhood heart disease in primary care	<p>Develop a Primary Health Care Childhood Heart Disease Strategy incorporating General Practitioners, Registered Nurses and Aboriginal and Torres Strait Islander Health Workers on the needs of patients of all ages with childhood heart disease and their families.</p> <p>Deliver education and training programs to support primary care and rural health professionals with a focus on referral pathways and the importance of communication between specialist and General Practitioner.</p> <p>Consider use of existing chronic disease management mechanisms such as GP Management Plans and Team Care Arrangements and Mental Health Plans to ensure access to appropriate support and holistic care.</p>	Peak Bodies TBC
2.5 Disseminate best practice information and approaches throughout services in Australia	<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>	CHAANZ and relevant Peak Bodies TBC

PRIORITY 2: SUPPORTIVE COMMUNITIES

FOCUS AREA 3: AWARENESS AND EDUCATION

For the purpose of this Action Plan,

Awareness and Education embraces the range of evidence-based communications, information and learning approaches often utilised 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.

The awareness and education challenges around childhood heart disease are quite complex and therefore responses are required to be comprehensive and multifaceted.

This Action Plan recognises the importance to those impacted by childhood heart disease of timely and readily available access to evidence-based, quality information and support. It also highlights the importance of the broader community being aware of the condition, its lifelong nature and the potential implications for those with the condition, their families and others in their life.

Greater awareness and education about childhood heart disease is critical to ensure that everyone affected by childhood heart disease has access to the right information and support across the life course and is supported to achieve optimal quality of life.

Although there is information and support currently available, these may not be evidence-based or appropriately targeted and, like 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.

Work in this area will be undertaken with four specific target groups in mind

1. Those impacted by childhood heart disease
 - The individual;
 - Their families and carers
2. Health professionals
3. The wider community, for example, education and employment settings, common services such as housing, insurance and finance.

Those impacted by childhood heart disease – the individual

Increasingly young people with childhood heart disease are surviving into adulthood. However, many drop out of the health system, unsure of how to access clinical care or having limited access to the services they feel they need or simply unaware that their condition requires lifelong care (CHAANZ, 2018). Therefore, increased awareness and education, incorporating information about and the availability of simple clinical pathways is critical, as is the motivation and confidence to act. Research has shown that increased referrals to appropriate CHD centres in line with clinical guidelines reduced mortality in adults with CHD (Mylotte et al., 2014).

Young people transitioning from the paediatric to adult health care system and from childhood through adolescence to adulthood and older age require specific, tailored information and resources delivered through appropriate modes and which go beyond the health and medical into aspects that affect their economic, social and emotional wellbeing.

“CHD, and the shadow it may leave, is for life. Not always a life sentence, but a lifelong concern; not always needing direct assistance, but always needing support.”

David

Those impacted by childhood heart disease – their families and carers

The awareness and education needs of parents and carers does not stop at finding out about their loved one’s condition and how best to support them. It requires them to know about and to be confident to contact support services and to navigate the complicated health care system. It also requires them to be able to advocate for their child and to inform and educate the myriad of professionals and services they must interact with about childhood heart disease. People impacted by childhood heart disease need to access information and support in a timely manner, regardless of where they are located. The lack of investment and overall infrastructure in childhood heart disease (beyond surgical interventions and clinical heart care) leaves many families isolated, with no clear pathway of care to seek support for suspected neurodevelopmental delay, behavioral and mental health issues for their child. It is acknowledged that currently, parents need to be experts but finding information, navigating systems and accessing services is difficult.

Lack of support is an identified issue for bereaved families as well. Families often feel isolated in the following weeks and months at home. 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 childhood heart disease.

The lived experience of families supports the research that shows that parental information on childhood heart disease is insufficient, and further suggests a link between information

dissemination and satisfaction with care (Lawoko, 2007). This is particularly the case for Aboriginal and Torres Strait Island families where differences in health literacy and English literacy may compound communication challenges (Wark et al., 2013, Mitchell et al., 2018).

“We didn’t get any more information – just told that we needed to come in for regular check-ups, and that he would go ‘blue’ from time to time when he cried.”

Parent feedback (Leggat, 2011)

Health and non-health professionals

Lack of awareness of childhood heart disease is not exclusively a consumer issue. Health professionals need to be versed in how to care for, refer and ensure they facilitate the best possible pathways to support desirable outcomes for people with childhood heart disease, including a holistic approach to their care. Health professionals have a role in communicating effectively with patients and their families and carers about childhood heart disease and to provide them with appropriate information and know how. They also have a role in creating an environment in which those impacted by childhood heart disease feel able to ask questions and discuss issues. Workforce capacity building for health professionals is considered in Focus Area 1: Standards of Care and explored further Focus Area 2: Infrastructure of this Action Plan.

Professionals in different sectors and settings have an important role to play in childhood heart disease care, including responding to emergency situations and promoting health and quality of life across the life course. Greater community awareness and knowledge has additional benefits such as ensuring that adults with childhood heart disease are not discriminated against by employers.

The broader community

As with all chronic conditions, supporting people impacted by childhood heart disease takes a whole of community approach.

Recent research conducted by HeartKids showed that barely a quarter of people are aware of HeartKids (as the only peak consumer organisation focused on supporting people impacted by childhood heart disease), congenital heart disease and even less aware of its impact on individuals and families (GALKAL, 2018).

Multiple research papers from Australia and internationally highlight the need for education of families and patients, and the community more broadly, including community settings, such as schools, work places, financial institutions and sports and recreation groups to ensure information about support and required intervention is available to assist people to navigate the system (Chong et al., 2018a).

What will be different?

- People impacted directly by childhood heart disease 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 health care 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 childhood heart disease 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.

DRAFT

<p>3.2 Increase access to ongoing support, by funding inpatient and outpatient Community (Peer) Support team members</p>	<ul style="list-style-type: none"> • 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>Community/Peer Support team based in hospital and community settings to provide support to individuals and families working alongside the clinical team to:</p> <p>Provide face to face support to families ensuring connected care before and after hospital admissions</p> <ul style="list-style-type: none"> • Facilitate connection to HeartKids community, peers as mentors, parents as advocates • Provision of HeartKids programs and services - tailored to family/community need • Liaison between Clinical Nurse Coordinators (see Focus Area 5: Priority Populations in this Action Plan), health service providers and cardiac social workers 	<p>HeartKids Community/Peer Support Workers</p>
<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> <ul style="list-style-type: none"> • In conjunction with young people, through HeartKids Youth Advisory Councils develop 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>Fund ongoing projects that facilitate connection with peers, support personal growth, self-management and leadership opportunities.</p> <ul style="list-style-type: none"> • 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, with appropriate resourcing for Indigenous health and social workers. 	<p>HeartKids in collaboration with Youth Advisory Council and multidisciplinary research team</p>

<p>Supportive Community: 3.4 Deliver awareness and education for non-health professionals</p>	<p>Fund a community-based awareness campaign to improve understanding of childhood heart disease and its impact across the whole of lifespan.</p> <p>The awareness campaign will focus on:</p> <ul style="list-style-type: none"> • Recognition of condition • Impact on families • Seeking treatment and need for lifetime care <p>Deliver awareness, education and provide support to professionals working outside the health care 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>	<p>HeartKids in partnership with consumers and key stakeholders</p>
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FOCUS AREA 4: NEURODEVELOPMENTAL AND MENTAL HEALTH CARE

For the purposes of this Action Plan,

Mental health care is defined as integrated, specialised and evidence-based care that focuses on reducing mental health burden and optimising emotional and psychobiological wellbeing for infants, children, young people and adults with childhood heart disease 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 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 childhood heart disease, with an emphasis on early intervention, ongoing surveillance and specialised intervention, as indicated.

Survival for children with childhood heart disease has significantly increased with the majority surviving into adulthood. Worldwide attention is now being directed toward understanding the long term quality of life, and mental, cognitive and emotional health outcomes and their trajectory in this growing population.

People with childhood heart disease 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.

Childhood heart disease has a profound impact on the health and wellbeing of those affected, with far reaching effects that ripple through a family and across a lifetime (Kasparian et al., 2016a).

People with childhood heart disease have a greater risk of neurodevelopmental impairment and disability including developmental delay and other learning difficulties.

People of all ages with childhood heart disease are at greater risk of lower health-related quality of life compared with their healthy peers, particularly in relation to neurodevelopmental, psychosocial and behavioural outcomes (Kasparian et al., 2016c, Ingles et al., 2013, Wilson et al., 2015a, Steptoe et al., 2000, Kovacs et al., 2009, Chong et al., 2017). Families of people with childhood heart disease, including parents, carers and siblings, also experience significant hardship and suffering (Kasparian et al., 2016c). See Appendix 1: The size of the problem for further information on the psychological and neurodevelopmental morbidity of childhood heart disease.

The neurodevelopmental and psychosocial morbidity related to childhood heart disease 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.

Examples of best practice mental health care exist in Australia, although care is inconsistent across the country and access is highly problematic, particularly for priority population groups such as Aboriginal and Torres Strait Islander people, families living in rural and regional areas, and families experiencing financial hardship. Deficiencies in specialist cardiac mental health services and workforce shortages are limiting patient access and comprising quality of care.

Despite the health, social and economic benefits of early intervention, the provision of neurodevelopmental care is limited by a lack of resourcing. While neurodevelopmental clinics and programs have been established at several paediatric cardiac centres across Australia, not all centres have these services and in those that do, not all children have access to timely neurodevelopmental surveillance, screening and assessment. Further, little attention is paid to neurocognitive health in ACHD care and neurocognitive assessment is not offered as part of routine care.

Optimal care for childhood heart disease involves cardiac care that integrates neurodevelopmental and mental health care as 'core business' (Kasparian et al., 2016b, Denniss et al., (in press)). Early intervention can make a profound difference for families and influence a lifetime of outcomes for a child (Kasparian et al., 2016a).

An emphasis on quality of life across the life course – beginning before birth, through infancy to adulthood – is required, so that medical advances over the past two decades are not undermined by the absence of adequate neurodevelopmental and mental health care (Kasparian et al., 2016a).

"We need to be focusing on outcomes beyond just survival – we need to move to a focus on quality of life."

Clinician (Leggat, 2011)

What will be different?

- Australia's national Standards of Mental Health Care and Neurodevelopmental Care for Childhood Heart Disease will ensure that neurodevelopmental and mental health care is a core part of childhood heart disease care and services, and subsequently, people with childhood heart disease and their families will experience improved mental health and cognitive outcomes and quality of life.

- People with childhood heart disease will have access to best practice neurodevelopmental and mental health care with a strong focus on optimising quality of life.
- The health and wellbeing of people with childhood heart disease and their families will be supported by a holistic approach to care that places equal emphasis on both physical and mental health.
- People with childhood heart disease 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 childhood heart disease and their peers will narrow.

“We have the capacity to lead the world in transforming mental health outcomes for children and adults with congenital heart disease, but we must take action now.”

Associate Professor Nadine Kasparian, Mental health clinician and researcher

Recommended actions: Neurodevelopmental and mental health care

Action	Detail	Lead
4.1 Develop Standards of Mental Health Care for Childhood Heart Disease	<p>As part of the development of Standards of Care for Childhood Heart Disease (Focus Area 1 in this Action Plan), develop world-first national Standards of Mental Health Care for Childhood Heart Disease.</p> <p>The Standards of Mental Health Care for Childhood Heart Disease will:</p> <ul style="list-style-type: none"> • Build on existing good practice, including The Heart Centre for Children’s four-tier model of integrated mental health care. • 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. • Be person- and family-focused, and inclusive of parents, carers and siblings. <p>Explore potential for transferability of standards of mental health care to other chronic conditions, such as cancer, asthma, neuromuscular diseases, other congenital anomalies, and other critical or chronic illnesses with childhood-onset.</p>	Childhood Heart Disease Taskforce, in partnership with the Heart Centre for Children, The Sydney Children's Hospitals Network
4.2 Develop Standards of Neurodevelopmental Care for Childhood Heart Disease	<p>As part of the development of Standards of Care for Childhood Heart Disease (Focus Area 1 in this Action Plan), develop national Standards for Neurodevelopmental Care for Childhood Heart Disease.</p> <p>The Standards of Neurodevelopmental Care for Childhood Heart Disease will:</p> <ul style="list-style-type: none"> • Build on existing good practice, including Queensland Paediatric Cardiac Service (State of Queensland (Queensland Health), 2018), and draw on international and 	Childhood Heart Disease Taskforce, in partnership with Queensland Paediatric Cardiac Service including CHD LIFE Program.

	<p>Australian resources including the American Heart Association’s best practice recommendations (Marino et al., 2012b).</p> <ul style="list-style-type: none"> • Focus on early intervention and ongoing surveillance across the lifespan • Address surveillance, screening, evaluation, diagnosis, and management of developmental disorders or disabilities in the paediatric congenital heart disease population. 	
4.3 Establish clear models of care	<p>Build on Queensland Paediatric Cardiac Service’s existing best 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"> • Known risk – ie children undergoing cardiac surgery under 12 months of age, post transplantation and survivors of extracorporeal life support (ECLS) are at greatest risk. • Family-centred – services and supports should be provided close to home • Long-term – Surveillance, screening and assessment into adolescence at recommended age points is required as deficits may emerge over time. <p>Test developmentally supportive practices in intensive care that incorporates developmental care and infant and parent mental health. Draw on best practice models such as the Royal Children’s Hospital’s COCOON model.</p> <p>The models of care will inform development of the Standards for Neurodevelopmental Care for Childhood Heart Disease.</p>	<p>Queensland Paediatric Cardiac Service including CHD LIFE Program.</p> <p>Royal Children’s Hospital, Victoria</p>

FOCUS AREA 5: PRIORITY POPULATIONS

For the purposes of the Action Plan,

Priority Populations are defined as populations that are disproportionately affected by childhood heart disease. 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 childhood heart disease and a greater burden of disease, resulting in inequitable health outcomes (Australian Health Ministers' Advisory Council, 2017).

Introduction

There is an urgent need to focus on reducing deaths and ill-health from childhood heart disease in priority populations.

Childhood heart disease impacts all Australians, but some populations are disproportionately affected. Greater investment and sustained efforts are required to positively advantage priority populations and overcome current inequities in health outcomes.

The overarching priority population of this Action Plan is people affected by childhood heart disease. Within this group, a number of priority populations have been identified (Box 2) based on extensive consultation with experts and the CHD community, and in response to evidence around the greatest need and potential for impact. Box 2 below.

Box 2: Priority Populations for Australia's National Action Plan for Childhood Heart Disease

The overarching priority population of this Action Plan is people affected by childhood heart disease. Within this group, priority populations include, but are not limited to:

- Aboriginal and Torres Strait Islander people
- Adolescents and young adults aged 15-24 years who are transitioning from paediatric to adult cardiac health services
- People living in remote, or rural and regional locations.

The approaches outlined in all priority areas of this Action Plan apply equally for priority populations as they do for the wider population. However, given the disproportionate burden of childhood heart disease experienced by priority populations, targeted action is essential to ensure that they receive access to quality, safe health care and relevant information and support. Action to improve health outcomes of priority populations will align with the renewed commitment to Closing the Gap and the new RHD Strategy recently announced by the Australian Government.

Aboriginal and Torres Strait Islander people

Both congenital heart disease and rheumatic heart disease are more common among Aboriginal and Torres Strait Islander people compared with non-Indigenous Australians, and Indigenous Australians experience worse outcomes than non-Indigenous Australians (AIHW, 2017a). This disparity is particularly stark with rheumatic heart disease.

A broad variety of cultural and systemic factors are barriers to early effective treatment among Indigenous Australians, many of which are exacerbated by remoteness (Ilton et al., 2014, Aroney et al., 2008, Australian Institute of Health and Welfare, 2016). Lack of culturally appropriate health care services, few resources in Aboriginal and Torres Strait Islander languages and systemic racism within the health system compound these issues (Mitchell et al., 2018, Brown and Kritharides, 2017).

Over 40% of people with childhood heart disease have to travel over 200 kilometres for specialist treatment, and have significant out-of-pocket expenses of \$2,500-3,500 (CHAANZ, 2018).

What will be different?

- The health system at all levels is responsive to the specific needs of priority populations to effectively address childhood heart disease 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 is 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 partner in, and lead, the planning, design, evaluation and implementation of locally responsive and culturally appropriate services.
- The disparity in health outcomes due to childhood heart disease between Aboriginal and Torres Strait Islander people and non-Indigenous Australians is reduced.
- Technology broadens access to health services, including appropriate use of telehealth and digital health options.
- The capability of the health workforce is enhanced to meet current and future needs of Aboriginal and Torres Strait Islander people and other priority population groups.
- Community empowerment and targeted action improves local and population health outcomes for priority populations with childhood heart disease.

Recommended actions: Priority Populations

Action	Detail	Lead
5.1 Fund CHD Cardiac Coordinators in each jurisdiction	<p>Fund CHD Cardiac Coordinators in each jurisdiction to coordinate and case manage rural/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.</p> <ul style="list-style-type: none"> • Registered Nurse role, with the potential to train and convert to Nurse Practitioner where appropriate • Total FTE 6.0; WA, NT and Queensland 1.5FTE; SA, NSW/ACT, Victoria/Tasmania 0.5FTE. (NT positions based in Darwin and Alice Springs, WA Perth and Kimberley, Qld Cairns and Brisbane.) <p>Key Performance Indicators to include:</p> <ul style="list-style-type: none"> • Specialist outreach clinics provided, activity and Failure To Attend rates • Coordination of surgery and interventional procedures • Aboriginal and Torres Strait Islander and total patients seen • Development of care pathways, including transition pathway • Successful transition to adult cardiac care • Education provided to young people with CHD • Education provided to rural/remote health staff • Annual report. 	Federal Government
5.2 Fund cardiac sonographers and a portable echo	<p>Fund cardiac sonographers and a portable echo machine in each jurisdiction. Rural/remote access to echocardiography is a priority. Echo is essential to CHD services and to the RHD program. In particular, access to echo for young people living with RHD is needed for monitoring disease progression, assessing the duration of preventative treatment and</p>	Federal Government

<p>machine in each jurisdiction</p>	<p>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 (RHD Australia, 2012). 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.</p> <ul style="list-style-type: none"> • Cardiac sonographers total 4.5FTE; WA, NT and Queensland 1.0FTE; SA, NSW/ACT, Victoria/Tasmania 0.5FTE • 8 portable echo machines 	
<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 outreach services where significant gaps exist 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.</p> <p>Utilise and expand telemedicine where appropriate.</p>	<p>CSANZ Federal Government</p>
<p>5.4 Establish models of care for the transition of care from paediatric to adult cardiac health services</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 addresses common reasons for ‘loss to follow up’ including insufficient patient/carer preparation, inadequate service integration and limited access to adult CHD specialists (Gurvitz and Saidi, 2014, Williams, 2015), and aims to reduce loss to follow up rates and re-engage adults lost to care.</p> <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>Royal Children’s Hospital</p>

<p>5.5 Investigate options for reducing barriers to accessing care and supporting CHD families</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> <p>Review disability and carer payments to ensure children and adults with congenital heart disease are not discriminated within the funding models. Current National Disability Insurance Scheme (NDIS) eligibility criteria excludes childhood heart disease as a medical condition despite the significant disability associated with chronic conditions including childhood heart disease.</p> <p>Provide support for people with childhood heart disease and their families to access and navigate existing services and government support including Carers Allowance, NDIS and Centrelink.</p>	<p>Relevant Federal Government Agencies TBC</p>
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PRIORITY 3: RESEARCH

FOCUS AREA 6: RESEARCH PRIORITIES

Australia has world-class researchers and is well placed to lead and develop childhood heart disease research on an international scale. These research priorities focus on congenital heart disease, with other research funding avenues identified for addressing acquired childhood heart disease including rheumatic heart disease (RHD).

Research and surveillance are essential to strengthening evidence-based practice for the diagnosis and management of congenital heart disease; identifying ways of preventing or curing congenital heart disease; 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.

As part of the HeartKids Conquering Childhood Heart Disease Research Mission, the Australian Government has provided an initial investment of \$15 million which will offer significant opportunities to make immediate and longer-term improvements in health outcomes for people living with congenital heart disease and their families.

The HeartKids Conquering Childhood Heart Disease Research Mission will initiate and accelerate research breakthroughs that change lives, gaining years of life by informing and optimising diagnosis and treatment.

The Action Plan and the accompanying *'HeartKids Conquering Childhood Heart Disease Mission' Research Framework* will accelerate and translate research into improved 'whole of life' care, in identified priority areas where Australia is, or can rapidly become, internationally competitive.

Investment will generate evidence to inform clinical practice and improve health care systems, promote new clinical trial activity, and provide greater access to innovative, cost-effective, personalised and accelerated interventions for infants, children, adolescents and adults with congenital heart disease.

The research guidelines outlined below, relate to the CHD Mission synergistically align with those of the MRFF:

- **Collaboration** both domestically and internationally to attract both talent and investment dollars.
- **International competitiveness** – invest in research that Australia is regarded internationally as a leader.
- Positioning Australia as an **incubator** for world-class research in emerging technologies and novel treatments.
- Funding research that **translates outcomes to patients in accelerated timeline** of 10 years to five years

- Research must have a **high degree of health consumer engagement** and meaningful participation.

HeartKids Ltd is currently developing Research principles as a key component of its Research Governance Framework. This document will be available as part of the final submission, as it is being developed concurrently. It is anticipated that these will reflect similar principles to the MRFF.

Current Collaborations are strong

Australian CHD specialist clinicians and researchers represent a strongly collaborative group with an established international reputation. Indeed, over the course of developing this Action Plan there has been substantial collaboration. The Conquering Childhood Heart Disease Mission builds and strengthens world leading collaborations developed through the Congenital Heart Alliance of Australia and New Zealand (CHAANZ) and the Australia and New Zealand Fontan Registry. Both programs highlight the collaborative and forward-looking approach of the childhood heart disease community and their willingness to work with health consumers on a range of internationally recognised projects.

Working closely with HeartKids, the national support organisation for the CHD patient and family community, these researchers and clinicians are uniquely well positioned to gain years of improved quality life for patients by innovating and optimising diagnosis, management and quality of care.

Currently HeartKids works with the following Research Organisations:

- Australian Centre for Heart Health
- Royal Princess Alfred Hospital
- The University of NSW
- Murdoch Children's Research Institute
- Victor Change Cardiac Research Institute
- Federation University
- Australia Grace Centre for Newborn Intensive Care,
- Westmead Children's Hospital
- The Sydney Children's Hospitals Network
- The Royal Children's Hospital Melbourne
- The University of Queensland
- Centenary Institute
- The University of Western Australia
- Lady Cilento Children's Hospital
- The University of Sydney
- Fontan Registry
- Congenital Heart Alliance of Australia And New Zealand

To translate research breakthroughs into improved 'whole of life' care, research priorities will be developed to 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.

What will be different?

- Evidence will inform clinical Standards of Care, 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 and 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.
- Identification of new strategies to remove barriers to adolescent and young adult clinical care helping close the gap on this identified Action Plan priority.
- Sharing best practice across Australia and internationally to increase engagement and improve outcomes for this vulnerable patient cohort.
- Purposeful collaboration with researchers across health services and institutions, and with the private sector.
- An identified model of monitoring and surveillance performance going forward

Recommended Research Priorities

Action	Detail
<p>6.1 Longitudinal Impact and Informed Practice with the Bi-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 health care. 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>Little is known about the economic burden of congenital heart disease. 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 and to update this report every two years.</p> <p>Accelerated implementation of the pilot CHAANZ Bi-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 following research priorities, enabling research collaboration, leveraging investment in the sector, and empowering patient and family interactions with the registry.</p>

	<p>Key deliverables</p> <ul style="list-style-type: none"> a) to optimise transition of care to allow “whole of life” management in the environment where young adults with CHD will have the best outcomes; b) to document variations in care and to allow optimal systems planning to build capacity in health services; and c) to facilitate research to understand Australian-relevant epidemiology and to determine effective interventions, and to establish databases to allow recruitment to clinical trials for novel interventions. This has the potential benefit to enable interface between patients and registry. d) Economic model of burden of disease both direct and indirect cost to be used to monitor performance of the Action Plan
<p>6.2 Preventing Neurodevelopmental and Mental Health Complications</p>	<p>Children who undergo major cardiac surgery as part of their treatment of CHD are at a major risk (up to 20%) of developing a neurodevelopmental disability (NDD) including developmental delay, motor and cognitive impairment and later school-based issues. 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, by prospectively assessing and screening neurodevelopmental abilities from birth, 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, 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> <ul style="list-style-type: none"> a) collect and analyse longitudinal data on neurodevelopmental impacts on children, and the mental health of patients and family members;

	<ul style="list-style-type: none"> b) determine appropriate approaches, either through genetic screening or identified clinical and social observations, for identifying those children at risk; and c) develop 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.
<p>6.3 Understanding CHD Genetics to Provide Personalised Medicine and Support for those Impacted by CHD - National Neonatal CHD Cohort Study</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:</p> <ul style="list-style-type: none"> a) identify the causes of structural heart disease in neonates and children and provide information regarding recurrent risks in further pregnancies and offspring of affected individuals; It is recommended this focus initially on foetuses with a diagnosis of an important single ventricle problem. b) develop clinical management and counselling strategies that are personalised according to the individual's genotype with expected benefits in perioperative management, drug dosing, prognostication and genetic counselling. 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, and c) Identify epigenetic mechanisms through which environmental factors exert their influence as a prelude to establishing preventative strategies. <p>Key Deliverables</p> <ul style="list-style-type: none"> a) Assess the clinical and psychological value of providing NATA (National Association of Testing Authorities) accredited whole genome sequencing and advanced genetic advice to families with multiple affected members, but who currently do not have access to clinical genetics services. This service will be created through establishment of multidisciplinary 'Cardiac Genetic Clinics' in Children's Hospitals and complement existing proposals intended to demonstrate the utility of genomic information in cardiovascular diseases; b) 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

	<p>pathways, defining risk and individualising care. This information would be available for early cardiac management, focused neurodevelopmental care and pharmacogenomic application.</p> <p>c) Build on the significant expertise of Australian Genomics Health Alliance and developmental biology researchers paired with clinicians to identify the most effective and most cost-efficient way of bringing the 'new genetics' to patients and families with CHD to prevent and treat CHD.</p> <p>d) The availability of prospective data on the 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.</p>
<p>6.4 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. 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. 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> <p>a) determine the benefits of tailored exercise and activity programs on improved exercise tolerance, muscle strength, activity levels and quality of life;</p> <p>b) define the mechanisms of how exercise and increases in activity benefit heart and physiological functions and adaptations that occur after cardiac rehabilitation;</p> <p>c) design and test models for sustainability and compliance of exercise programs;</p> <p>d) develop guidelines for personalised exercise and activity programs relevant to the large range of structural variations in CHD, in coordination with CSANZ; and</p>

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| | e) 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. |
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FOCUS AREA 7: SURVEILLANCE

For the purposes of this Action Plan,

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 (World Health Organization, 2018).

Effective surveillance of childhood heart disease is needed to understand and monitor the epidemiology of the disease in order to set priorities and guide health policy and strategies. Importantly, it will also be used to monitor the impact of this Action Plan and progress towards its Goal.

As recommended in this Action Plan, a Childhood Heart Disease Task Force should be established to oversee the design, development and implementation of National Childhood Heart Disease Standards of Care. It is proposed that the Task Force also assume the accountability for the Surveillance and Monitoring of the Action Plan. It is recommended that the Task Force work closely with the Report's key collaborators, HeartKids Limited, the Congenital Heart Alliance of Australia and New Zealand (CHAANZ) as the CHD Registry managers and the Paediatric and Congenital Council of the Cardiac Society of Australia and New Zealand (CSANZ).

The Task Force's terms of reference would therefore include:

- Annual collection of data
- Consultation with State and Territory Health Departments, health consumers and other stakeholders
- Provide advice to AIHW and others on appropriate data sets to going forward to measure impact
- Preparation of a report to the COAG Health Council.

Recommended Actions: Surveillance

Action	Detail	Lead
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.	Congenital Heart Alliance of Australia and New Zealand (CHAANZ)
7.2 Continued funding of the National CHD Survey	<p>Continue funding the National CHD Survey to collect data from people with childhood heart disease, including their health care use and experiences, and the health, social and economic impact of living with CHD.</p> <p>Survey to be administered annually to monitor change over time.</p>	CHAANZ
7.3 Develop an annual report card to monitor progress against this Action Plan	Develop an annual report card to monitor the field's progress against the Goal of this Action Plan. See 'Monitoring Progress' section of this Action Plan.	HeartKids and CHAANZ

MONITORING PROGRESS

A critical element of the Action Plan's success is appropriate ongoing investment by stakeholders including Australian and State and Territory Governments in the evaluation and monitoring of the Plan's implementation.

Consumers during consultation were very concerned, following the stalled 2011 White Paper, that accountability for monitoring improvements over time be clearly assigned.

Further given the rapidly evolving nature of health care in Australia, the emerging challenges of childhood heart disease in adulthood and the significant concerns regarding infrastructure planning, many consumers expressed a desire for an annual independent assessment of progress.

In this regard it is proposed to fund the Congenital Heart Alliance of Australia and New Zealand (CHAANZ) and HeartKids to complete and publish an Action Plan Annual Report Card (Action 7.3 in this Action Plan).

CHAANZ will use existing infrastructure now deployed in the CHD Registry and National Survey of CHD Patients to track over time the Action Plan recommendations. Impact of the Action Plan is proposed to be undertaken via the CHAANZ Registry Data and the National CHD Survey and through focus group interviews of this Plan's key stakeholders.

In addition, it is proposed to also conduct three Economic Impact Reports in year's one, three and five (post report release) to measure any improvements to the economic and productivity burden of the disease on the Australian Community.

To measure progress, the scorecard will include data such as:

Progress on recommended actions, including:

- Number of actions funded (full versus partial funding)
- Number of actions initiated at Australian, State and Territory Governments and key stakeholders including HeartKids and the Cardiac Society of Australia New Zealand (CSANZ)
- Number of actions completed
- Number of actions completed according to timeframe, budget and scope

Outputs of the Action Plan, including:

- Changes to health care services and family support, such as new regional clinics, CHD education portal;
- Key strategic documents, such as Standards of Care for Childhood Heart Disease, Workforce Plan and Infrastructure Plan;
- Contributions to the evidence base, such as stronger evidence of effective interventions to improve childhood heart disease care.

The recommended baseline measures of success are:

- Improved quality of life and mental health among people with childhood heart disease and their families
- Improved neurodevelopmental outcomes for people with childhood heart disease
- Increased access to services, including lead and lag indicators for surgery and allied health services addressing mental health and psychosocial impacts
- Reduced childhood heart disease-related deaths and premature deaths (Indigenous and non-Indigenous)
- Reduce the incidence of rheumatic heart disease
- Reduce the need for cardiac surgery for rheumatic heart disease by early diagnosis and improved self-management strategies
- Improved loss to follow-up rates for adolescents and young adults with childhood heart disease when transitioning from paediatric to adult health services
- Reduced social and economic burden of childhood heart disease to individuals, families, the healthcare system and government, including impact on educational institutions and workforce productivity.

Additional measures will be developed to monitor progress against key initiatives and areas of work.

ACKNOWLEDGEMENTS

Development of the Childhood Heart Disease National Action Plan 2018 was led by HeartKids Ltd 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 Plan, including people with childhood heart disease, health professionals, key medical and cardiac organisations, the research community and the Australian Government and State and Territory Governments.

HeartKids sincerely thank the members of the Childhood Heart Disease National Action Plan Steering Committee and Expert Working Groups including the hard-working 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 childhood heart disease is greatly appreciated.

Steering Committee

- Ms Jan McClelland AM, Chairperson, HeartKids
- Associate Professor Gary Sholler, Director Cardiac Services, Sydney Children's Hospital Network
- Associate Professor Michael Cheung, Director of Cardiology, Royal Children's Hospital Melbourne
- Associate Professor Robert Justo, Director Paediatric Cardiology, Queensland Health
- Professor David Celermajor, CHAANZ Steering Committee; Board Director, HeartKids; Royal Prince Alfred Hospital; University of Sydney
- Professor Yves d'Udekem, Deputy Director, Department of Cardiac Surgery, Royal Children's Hospital Melbourne
- Dr Gavin Wheaton, Medical Director, Women's and Children's Hospital Adelaide
- Dr James Ramsay, Paediatric Cardiologist, Perth Children's Hospital
- Dr Andrew Kelly, Medical Unit Head Cardiology, 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
- Dr Lisa Selbie, Chairperson Research Advisory Committee, 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 Simone Wright, Consumer representative, Parent of a CHD child
- Ms Michelle Pendrick, Consumer representative, Adult with CHD
- Mr Mark Brooke, Chief Executive Officer, HeartKids

Expert Working Groups

Research and Infrastructure:

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- Dr Lisa Selbie, Chairperson Research Advisory Committee, HeartKids
- Associate Professor Enzo Porrello, Co-Director, Melbourne Children's Centre for Cardiovascular Genomics and Regenerative Medicine
- Associate Professor Nadine Kasparian, Head of Psychology, Heart Centre for Children, Sydney Children's Health Network
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- Professor Richard Harvey, Co-Deputy Director, Victor Chang Cardiac Research Institute
- Professor Jonathan Carapetis, Director, Telethon Kids Institute
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- Dr Claire O'Donnell, Paediatric Cardiologist, Starship Children's Hospital Auckland
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- Mr Dwayne & Mrs Robyn Broome, Consumer representative, Adult with CHD
- Ms Michelle Pendrick, Consumer representative, Adult with CHD.

Standards of Care:

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CHD & Co-morbidities:

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- Associate Professor Gary Sholler, Director Cardiac Services, Sydney Children's Hospital Network
- Associate Professor Robert Justo, Director Paediatric Cardiology, Queensland Health
- Professor Alun Jackson, Director, Australian Centre for Heart Health
- Professor Nadia Badawi, Medical Director, Grace Centre for Newborn Intensive Care, Westmead Children's Hospital Sydney
- Dr Gillian Blue, Genetic Counsellor, Sydney Children's Hospital Network
- Dr Julia Charlton, Neonatologist, Royal Children's Hospital Melbourne
- Dr James Ramsay, Paediatric Cardiologist, Perth Children's Hospital
- Ms Karen Eagleson, Clinical Nurse Consultant Paediatric Cardiology, Queensland Health
- Ms Michelle Pendrick, Consumer representative, Adult with CHD
- Ms Simone Wright, Consumer representative, Parent of a CHD child.

Creating Supportive Community Environments:

- Dr Gavin Wheaton (Chair), Medical Director, Women's and Children's Hospital Adelaide
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- Professor Jonathan Carapetis, Director, Telethon Kids Institute
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- Dr James Ramsay, Paediatric Cardiologist, Perth Children's Hospital
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- Ms Cecilia Donovan, State Manager WA HeartKids; Consumer representative, Parent of a CHD child
- Ms Holly Williams, State Manager QLD HeartKids; Consumer representative, Adult with CHD and Parent of a CHD child
- Mr Dwayne & Mrs Robyn Broome, Consumer representative, Adult with CHD
- Ms Michelle Pendrick, Consumer representative, Adult with CHD.

Secretariat

- Mr Mark Brooke, Chief Executive Officer, HeartKids (until September 2018)
- Mr Rob Daly, Acting Chief Executive Officer, HeartKids (from September 2018)
- Ms Paula Murray, General Manager Programs, Research and Innovation, HeartKids
- Ms Maree Davidson AM, Davidson Consulting (Strategy and Facilitator)
- Ms Rebecca Zosel, Zosel Consulting (Principal Writer)

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APPENDIX 1: THE SIZE OF THE PROBLEM

PREVALENCE

Around 65,000 - 90,000 Australian children and adults live with childhood heart disease.

Congenital heart defects are the most common types of birth defects, affecting 1 in every 100 babies.

Every day in Australia, eight babies are born with congenital heart disease; worldwide there are about 1.35 million babies with heart disease each year (van der Linde et al., 2011).

In Australia, 2,400 – 3,000 babies are born each year with a form of congenital heart disease – approximately 8-10 cases per 1,000 live births.

The most commonly reported congenital heart conditions in Australia include ventricular septal defect (630 cases), patent ductal arteriosus (406), atrial septal defect (402), pulmonary stenosis (134) and transposition of the great vessels (103) (AIHW, 2017a).

The prevalence of congenital heart disease is predicted to increase, particularly the adult population.

Around 65,000 - 90,000 children and adults are currently living with a childhood heart condition in Australia. A lack of definitive research on the burden of disease suggest the number could be far higher. This includes around 32,000 children under 18 years of age, and over 32,000 adults who have lived with a heart condition since childhood. For the first time in history, adults living with childhood heart disease outnumber children with the disease.

As Australian birth rates increase and enhanced medical care and technology continue to improve survival rates following medical interventions, the prevalence of congenital heart disease is predicted to increase, particularly the adult population which is predicted to increase annually by 5% (Leggat, 2011).

Both congenital heart disease and rheumatic heart disease are more common among Aboriginal and Torres Strait Islander people compared with non-Indigenous Australians, and Indigenous Australians experience worse outcomes than non-Indigenous Australians (AIHW, 2017a).

Understanding the true prevalence of childhood heart disease in Australia is challenging. Prevalence data is often extrapolated from the Netherlands national CHD registry and applied to the Australian population. The most recent national data on the prevalence of congenital heart disease is from the 2003 Australian Congenital Anomalies Monitoring System. In time, the recently launched Congenital Heart Disease Registry will provide

essential information on childhood heart disease in Australia. Data about the prevalence of childhood heart disease and burden of disease in Australia is needed to provide an evidence-based platform for setting priorities and guiding health policy and strategies.

MORTALITY

Congenital heart disease is a leading cause of death of Australian babies (AIHW, 2018).

Congenital heart disease is a leading cause of death of Australian babies. Among infants, perinatal and congenital conditions caused most deaths (77% of deaths), as shown in Figure 5 (AIHW, 2018).

Figure 5: Leading underlying causes of death, by age group, 2014–2016 (AIHW, 2018)

AGE GROUP	1 ST	2 ND	3 RD	4 TH	5 TH
UNDER 1	Perinatal and congenital conditions	Other ill-defined causes	SIDS	Spinal muscular atrophy	Accidental threats to breathing
1-14	Land transport accidents	Perinatal and congenital conditions	Accidental drowning and submersion	Brain cancer	Other ill-defined causes
15-24	Suicide	Land transport accidents	Accidental poisoning	Assault	Other ill-defined causes
25-44	Suicide	Accidental poisoning	Land transport accidents	Coronary heart disease	Other ill-defined causes
45-64	Coronary heart disease	Lung cancer	Suicide	Breast cancer	Colorectal cancer
65-74	Lung cancer	Coronary heart disease	COPD	Cerebrovascular disease	Colorectal cancer
75-84	Coronary heart disease	Dementia and Alzheimer disease	Cerebrovascular disease	Lung cancer	COPD
85 AND OVER	Coronary heart disease	Dementia and Alzheimer disease	Cerebrovascular disease	COPD	Heart failure

Historically, survival of congenital heart disease patients with significant lesions beyond early childhood was limited, with less than one-third of infants being expected to survive into adulthood. The number of deaths from childhood heart disease in Australia has significantly declined over the last three decades, particularly in children. The majority (>95%) of people with childhood heart disease now survive into adulthood (Le Gloan et al., 2011, Khairy et al., 2010), however life expectancy continues to be below normal (Winlaw, 2007).

The majority (>95%) of people with childhood heart disease now survive into adulthood, however life expectancy continues to be below normal.

Inequalities in outcomes for young Aboriginal and Torres Strait Islander with rheumatic heart disease are particularly pronounced. Young Indigenous people (<35 years) are 122 times more likely to have RHD than their non-Indigenous peers (Australian Institute of Health and Welfare, 2013). Between 2011–2015 94% of people diagnosed with acute rheumatic fever identified as Indigenous and 55% of them were aged 0 – 14 years (Australian Institute of Health and Welfare, 2017). On average, Aboriginal people with rheumatic heart disease die at 41 years of age (Davies et al., 2014).

MORBIDITY

Childhood heart disease is a major global health burden.

Despite remarkable medical success in recent years, childhood heart disease continues to be a major global health burden (Celermajer et al., 2016).

There is clear evidence that people of all ages with congenital heart disease are at greater risk of lower health-related quality of life compared with their healthy peers, particularly in relation to neurodevelopmental, psychosocial and behavioural outcomes (Kasparian et al., 2016c, Ingles et al., 2013, Wilson et al., 2015a, Steptoe et al., 2000, Kovacs et al., 2009, Chong et al., 2017). Families of people with childhood heart disease, including parents, carers and siblings, also experience significant hardship and suffering (Kasparian et al., 2016c).

People with childhood heart disease 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.

Although medical advances have reduced paediatric mortality and extended life expectancy for those with childhood heart disease, psychological morbidity remains prevalent. Reported prevalence of anxiety is higher than depression, both of which have been generally found to exceed population norms, ranging from 15-33% in patients of all ages (Kasparian et al., 2016c, Ingles et al., 2013, Wilson et al., 2015a, Steptoe et al., 2000, Kovacs et al., 2009).

The presence of anxiety and depression is associated with a range of difficulties, including but not limited to, increased risk of major cardiovascular events, shorter event-free survival (Kourkouveli et al., 2015), lower adherence to cardiac care recommendations (Wray et al., 2013), and impaired health-related quality of life (HRQOL) (Steptoe et al., 2000). Higher

levels of cardiac events can be a major burden on Australia's health care system and levels of workforce participation and productivity.

Correlates of anxiety and depression include lower perceived health status (Kovacs et al., 2009), the presence of neurodevelopmental delay (Latal et al., 2009), greater social isolation (Kovacs et al., 2009), and poorer self-perception (Czosek et al., 2016). Approximately 30-80% of parents of children with complex congenital heart disease also report high psychological distress (Woolf-King et al., 2017).

Poorer parental mental health is associated with greater suicidal ideation, poorer physical health and greater healthcare usage, in addition to poorer family functioning and child emotional, cognitive, behavioural, social and health-related outcomes (Kasparian et al., 2016c).

Families of people with childhood heart disease including parents and carers experience significant hardship and suffering (Leggat, 2011, Chong et al., 2018b). Parents of children with CHD experience more anger and sadness than parents of healthy children (Cohn, 1996), more social problems and less leisure activities (Casey et al., 1996), greater feelings of distress and hopelessness (Lawoko and Soares, 2002), and higher levels of overall stress than parents with children with other chronic conditions (Goldberg et al., 1990, Uzark and Jones, 2003). Research shows that 1 in 3 mothers and fathers of infants with complex CHD report symptoms that meet clinical criteria for depression, and about 50% of parents report severe stress reactions consistent with a need for clinical care - up to one year after their baby's diagnosis (Kasparian NA et al., 2010).

These rates far exceed documented rates of perinatal depression and anxiety in the general community (Buist A and Bilszta J, 2006) and as healthcare providers, we often markedly underestimate the severity and potential consequences of these symptoms (Kasparian et al., 2016).

Our understanding of the psychology of adult CHD lags decades behind our knowledge of children's experiences. Some research has found that one in three adults with CHD report symptoms of anxiety or depression warranting intervention (Kovacs AH et al., 2009). The vast majority of these adults go untreated (Kasparian et al., 2016a).

People with childhood heart disease have a greater risk of neurodevelopmental impairment and disability including developmental delay and other learning difficulties.

Children who undergo open heart surgery, particularly in the first year of life are known to have a higher risk of impairment across multiple domains when compared with the general population including intellectual ability, behaviour, social interaction, communication and executive function (Marino et al., 2012a, Majnemer et al., 2006, Bellinger et al.). The underlying cause of these deficits is multifactorial, with complex interactions between genetic susceptibility, physiological consequences of the heart defect and circulation,

perioperative brain injury and psychosocial factors all contributing (Donofrio et al., 2011, Donofrio and Massaro, 2010).

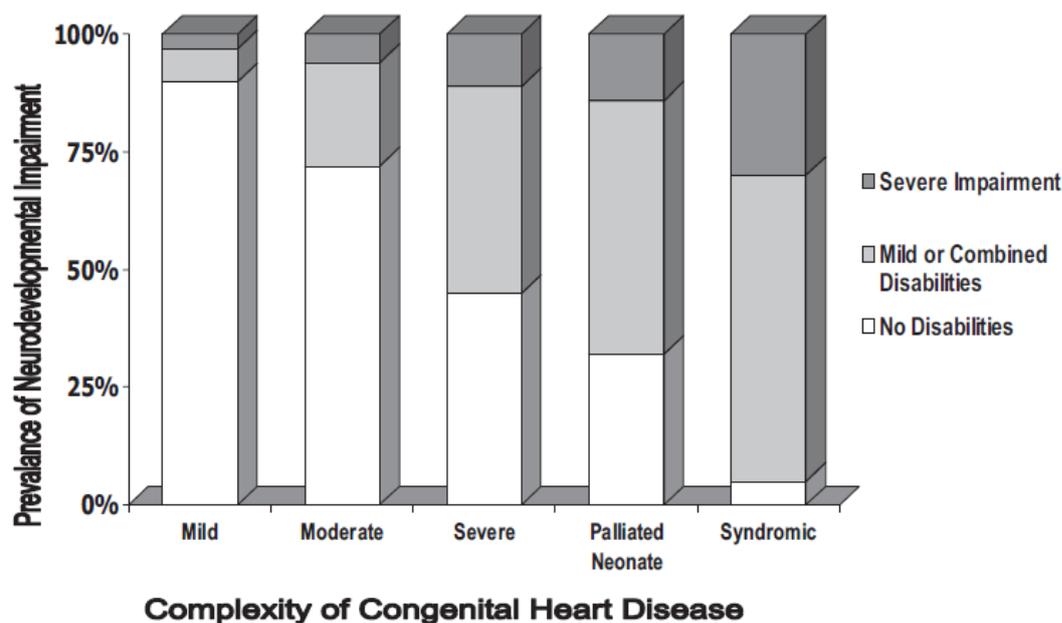
Children with complex congenital heart disease (including those who need surgical intervention in the first year of life and those with other, comorbid physical conditions) are at increased risk of neurodevelopmental impairment and disability – see Figure 6 (Marino et al., 2012b). The risk and severity of neurological impairment increases with greater CHD complexity, the presence of a genetic disorder or syndrome, and greater child and family psychological stress (Marino et al., 2012b).

During infancy, the most pronounced difficulties occur in motor functioning. By early childhood, neurological impairment is characterised by difficulties in fine and gross motor skills, speech and language, attention, executive functioning, emotion regulation and behaviour (Marino et al., 2012b).

Studies also show that children with complex CHD are also up to four times more likely to be diagnosed with attention deficit and hyperactivity disorder than the general population (Wilson et al., 2015b).

The neurodevelopmental and psychosocial morbidity related to childhood heart disease 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.

Figure 6: Neurodevelopmental outcomes in children with congenital heart disease (Marino et al., 2012b)



ECONOMIC IMPACT

The economic burden of childhood heart disease has not been well described, and it is difficult to estimate the total economic impact of childhood heart disease to the health care and education systems, governments, workforce and people affected by childhood heart disease.

Childhood heart disease poses a substantial economic burden on our health care system. Congenital heart disease is one of the leading causes of hospitalisation in the first year of life (AIHW, 2017b), and successful treatment requires highly specialized care.

Childhood heart disease requires extensive financial resources both in and out of the hospital, with costs directly related to the degree of disease complexity. A Hong Kong study calculated the annual cost of management of simple ACHD was USD 2638 versus complex ACHD was USD 6425 (Lee et al., 2016).

Surgery for childhood heart disease results in an enormous financial burden for the majority of families at a very vulnerable time (Raj et al., 2015). Significant out-of-pocket costs exist for patients and their families, including travel and accommodation for families required to travel to the designated CHD specialist centres. This burden is particularly high for families in rural and remote locations including Aboriginal and Torres Strait Islander families.

A recent study showed that over 40% of people with CHD have to travel over 200 kilometres for specialist treatment, and have significant out-of-pocket expenses of \$2,500-3,500 (CHAANZ, 2018).

The large and growing population of adults with childhood heart disease require lifelong and highly specialised medical care. This changing demographic and epidemiology of CHD has shifted a large portion of the burden of disease into the adult population, with little understanding of the cost to patients, the health system, and the community.

Social burdens of childhood heart disease including education, employment and insurability, which increase the societal costs of adult CHD, are now being recognised for adults living with CHD (Seckeler et al., 2016).

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