



The Cardiac Society of Australia and New Zealand

## Adult Congenital Heart Disease (ACHD) Recommendations for Standards of Care

Development of this Position Statement was co-ordinated by Prof David Celermajer, Drs Mugur Nicolae, Tim Hornung, Clare O'Donnell and members of the Paediatric and Congenital Council; Drs Robert Justo, Andrew Bullock, Michael Cheung, A/Prof Andrew Cochrane, Drs Tom Gentles, James Ramsay, A/Prof Gary Sholler and Dr Gavin Wheaton. No authors have any relevant Conflict of Interest to disclose.

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### SUMMARY

- The Cardiac Society of Australia and New Zealand (CSANZ) recognises that adult congenital heart disease (ACHD) is a relatively new and rapidly growing area of need, for which appropriate planning is required to establish and maintain adequate standards of care.
- There are likely now more adults (over 16 years) in Australia and New Zealand (ANZ) than children with CHD, and the numbers of ACHD patients are increasing annually, particularly those with complex disease.
- Recent data have emphasised that (i) without adequate planning, “loss to follow up” rates in transition from paediatric to adult congenital heart services are over 50% and that (ii) mortality rates are substantially higher for young adults lost to follow up, compared to those under regular review at a dedicated ACHD centre. A recent publication has also documented a mortality benefit for those young adults with ACHD cared for in specialist centres, compared to those who are not (Wray et al, Heart 2013; 99: 485-90).
- CSANZ wants to emphasise that a “whole of life” plan for the management of Congenital Heart Disease is appropriate, in which context the issues of excellence in paediatric cardiac services, excellence in adult CHD services and seamless transition of care arrangements should be available to patients with CHD across Australia and New Zealand.
- The two key areas requiring attention are:
  - i. Planning of Transition of Care from paediatric cardiac to adult CHD services.
  - ii. Establishing and maintaining Comprehensive and Regional ACHD services to serve the population needs of ANZ.

Guidelines addressing these two aspects are attached.

## Transition of Care Arrangements for Adolescents / Young Adults with Congenital Heart Disease

### PREAMBLE

Most children with Congenital Heart Disease are not “cured” by their paediatric interventions, rather many of them have residual haemodynamic and/or electrophysiologic abnormalities that require long term surveillance.

Transition of care from the Paediatric Cardiology to the Adult Congenital Heart Disease (ACHD) environment is difficult and challenging. The transition occurs from a “trusted” team to an unknown team in a different environment. International studies have suggested that even in the best models, up to 50% of children may not successfully transition and may be lost to long term follow up, with potentially catastrophic consequences for their health.

Well established ACHD units have the capacity to cater for not just their cardiac needs but ancillary medical needs, such as anaesthetic and/or obstetric support where necessary, and their psychosocial and non-medical needs also.

### SUGGESTED GUIDELINES

CSANZ believes that transition is best done by a close working relationship between the Paediatric Cardiac Team and the ACHD centre or specialist, with some responsibilities accruing to these entities, as well as some responsibilities belonging to the patient and family. CSANZ wishes to encourage a “whole of life” approach with a continuum of care. Transition should be *from* a paediatric cardiologist or paediatric cardiac service to a specific ACHD Specialist or to a recognised ACHD centre.

1. We recommend that each major Paediatric Cardiac Unit employ a Transition Nurse or health professional, at least 50% FTE, to oversee and ensure effective transition arrangements, at an individual patient level. The effectiveness of Transition to ACHD care should be a Key Performance Indicator for the Paediatric Cardiac Unit.

In general, the Paediatric Cardiac Team should raise the concept of transition of care at an appointment in early teenage years; and should provide the patient and their family with a paper or electronic copy of their final clinical evaluation and any relevant diagnostic tests (e.g. CXR, ECG, echocardiogram and/or exercise test, other forms of imaging). This “information pack” should also include ideally a copy of the actual operation reports from significant heart surgeries.

Where the local ACHD centre has an Information Brochure, they should be provided to patients and their families.

2. The plan for transition should be communicated to the patient’s normal GP.
3. The Paediatric Cardiac Team should clarify that the patient will continue to receive medical support from the Paediatric Cardiac Team until the patient has met the ACHD specialist/team for the first time.
4. The ACHD specialist or centre to whom the patient is referred should make appointments for the patients of the Paediatric Cardiologists when requested, including providing contact details for each patient to use during the time of transition; and to notify the Paediatric Cardiologist when the transition documentation has been received.
5. This service should attempt to make a reminder call to each transitioning patient in the week prior to their scheduled appointment and to have a mechanism to follow them up should they fail to attend.

## Centres for Adults with Congenital Heart Disease

Across Australia and New Zealand there should be:

- a) A Comprehensive Adult Congenital Heart (CACH) Service in those regions serving populations of  $\geq 2$ -3 million people. Where the population served is over 5 million people, consideration should be given to 2 CACH services; depending on local factors and geographies.
- b) A Regional Adult Congenital Heart (RACH) Service for regions which are at a substantial distance from a CACH service.
- c) If possible, CACH and RACH centres should develop an Information Brochure explaining their location, staff, contact details and purpose of follow up, for Paediatric Cardiac Units to distribute as part of easing the concerns of patients and families around the Transition process.

Comprehensive Adult Congenital Heart Centres (CACH) should have established links to the regional Paediatric Cardiac centres. They should hold regular Multidisciplinary Case Conferences, to optimise decision making. CACH centres should comprise:

- a) At least 2 ACHD specialists, who are adult FRACP cardiologists who have spent at least 12 months of training in dedicated paediatric or adult congenital cardiology units; or paediatric cardiologists with specific experience in ACHD (acknowledging that no formal accreditation system for ACHD specialisation currently exists).
- b) At least 2 ACHD surgeons, who are FRACS-qualified paediatric cardiac surgeons with specific experience in ACHD.
- c) Access to the following services:
  - i. Cardiac MRI and a cardiologist or radiologist experienced in ACHD.
  - ii. Electrophysiology services with at least one EP specialist with ACHD expertise.
  - iii. An interventionist with experience in treatment of ACHD.
  - iv. An Intensive Care Unit with adequate capability for peri-procedural support.
  - v. A high risk/medical Obstetrics Unit that supports ACHD patients.
  - vi. A heart and heart/lung transplant program, in the same city.
- d) A Clinical Nurse Coordinator and a Nurse Educator.
- e) Access to Clinical Psychology, Genetic Counselling and Social Work support services.

Regional Adult Congenital Heart Services should have at least one ACHD cardiologist, as defined above; a Clinical Nurse Coordinator (at least part time); and established links to a CACH service.

Every young adult with repaired CHD should be seen at least once in an ACHD specialist centre, except those who have been discharged from follow up by their paediatric cardiologist.

Every young adult with unrepaired CHD should be seen regularly at an ACHD facility.

Every adult with non-simple CHD (where simple CHD is small or repaired ASD, VSD or PDA without residual haemodynamic abnormality; or mild pulmonary or aortic valve disease) should be seen regularly at an ACHD centre, with or without shared care by community-based specialists.