CHD and RHD: Promoting Long-Term Health

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Mission: Improve worldwide life-long outcomes in childhood-onset heart disease through empowering patient and family organizations

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- 40 RHD and CHD groups; 29 countries, >½ LMICs
- US 501c3

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Today's Talk

- **Focus on P/F group activities**
- CHD and RHD long-term issues
- Core educational messages
- Using CHD/RHD guidelines as resource

Not: *Clinical Care, Research Overview, Individual lesions*

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Will my child have a normal life?
Were you able to have children?
What risks do they face as they get older?
What kind of care will they need?

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RHD	CHD
Caused by untreated strep infection	Most cases unknown cause
Preventable	Unpreventable
Disease of poverty, LMICs	Affects all comers
Almost eradicated in HI countries	Common in HI countries
School age children/young adults	Present at birth

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Health Problems	CHD	RHD
Problems with heart valve(s)	X	X
Problems with other heart structures	X	
Risk of strep re-infection		X
Heart rhythm problems	X	X
Risk of stroke	X	X
Weakened heart function/heart failure	X	X
Heart infection (endocarditis)	X	X
Lung problems (pulmonary hypertension)	X	X
Problems with pregnancy	X	X
Genetic risk to children	X	?

Care maintenance is essential

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Barriers to Long-term Follow-up	CHD	RHD
Lack of local/regional providers	X	X
Financial barriers	X	X
Psychosocial issues/stigma	X	X
Lack of understanding of care needs	X	X
Perception of Cure	X	x

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They Say	We Hear
“Complete Surgical Repair”	
“Corrective Surgery”	“She’s cured”
“Complete Anatomic Repair”	“I’m fixed”
“Total Correction”	
“Reparative surgery”	
“What did you/she have?”	“I don’t have it anymore”

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Healing Little Hearts

Give a child a new life

HRUDAYA

Cure a little heart Foundation

putting the smile back

HRUDAYA

Cure a little heart

FOUNDATION

"My son was born with a heart problem, but he had surgery so he is fine now"

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Problem of Success

- Most post-OHS children do well throughout childhood
- Minimal/no limits

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1960 – 1990

"Your child is fixed"

"Go live a normal life"

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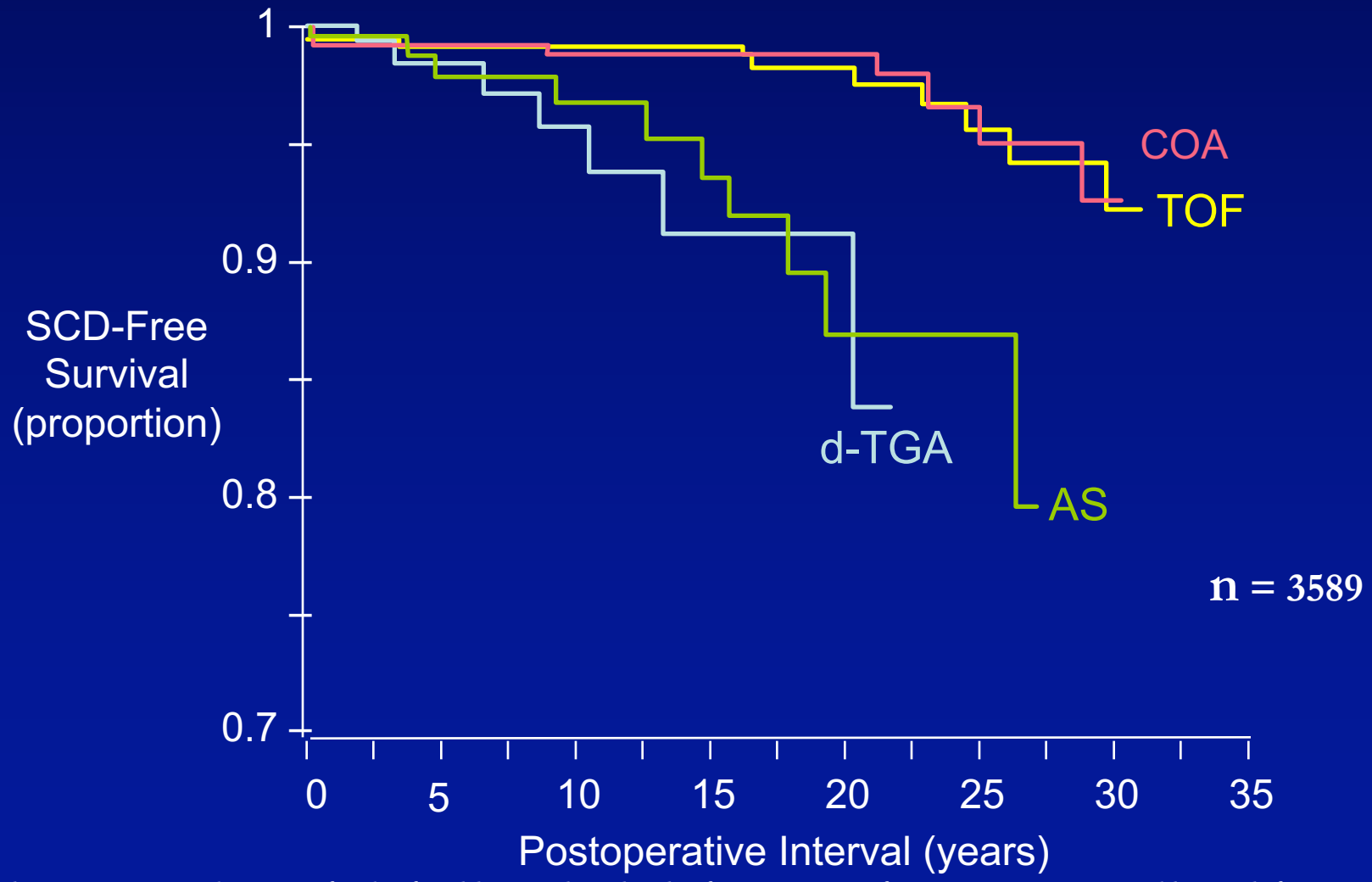
Probability of SCD-Free Survival After Surgical Treatment

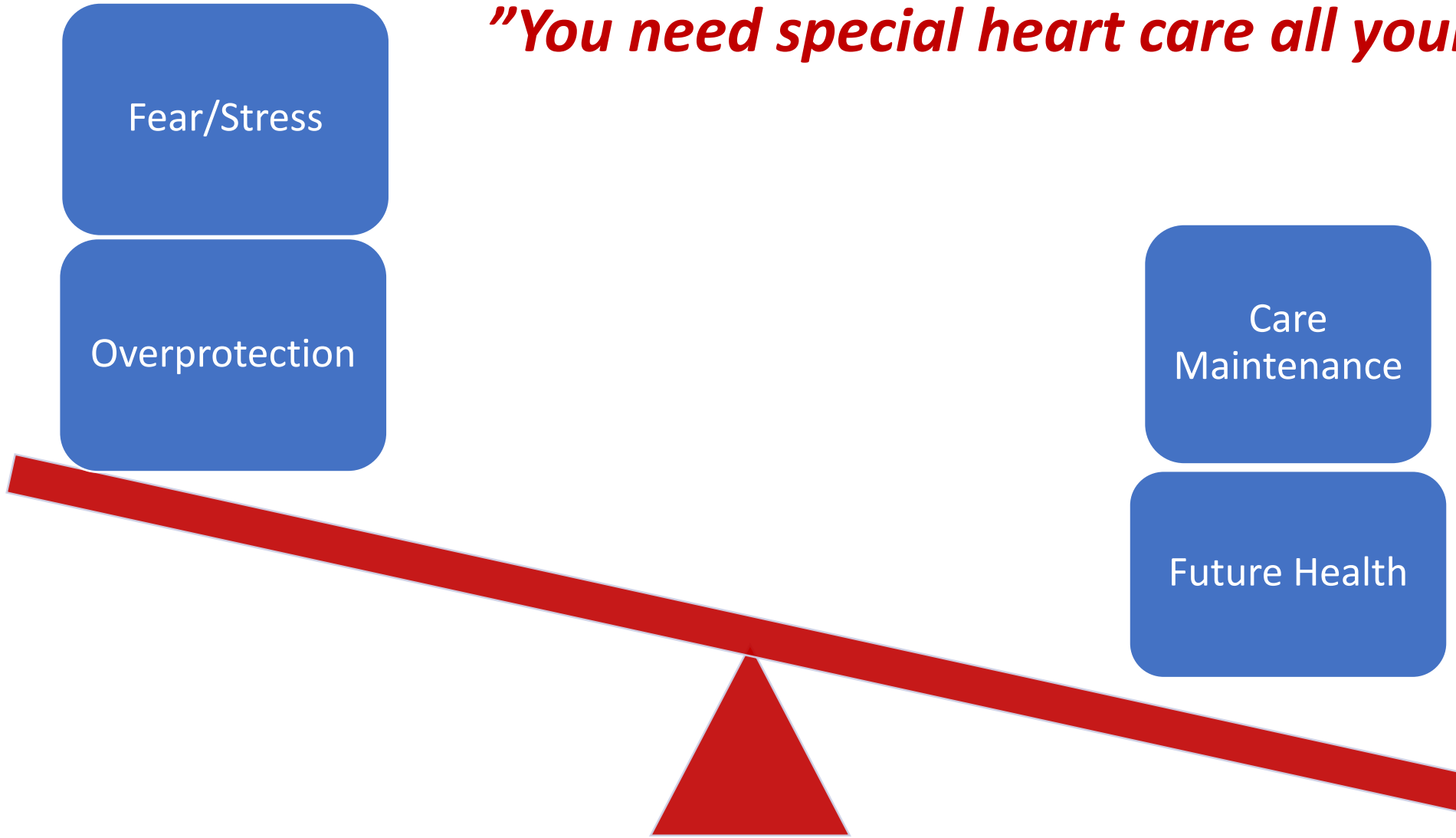


Silka et al. A population-based prospective evaluation of risk of sudden cardiac death after operation for common congenital heart defects *JACC.* 1998; 32: 24. 5-251

2000 – Present

"Your child can do very well but will never have a normal heart"

"You need special heart care all your life"



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Basic Educational Messages

- CHD (even post-repair) and RHD need life-long care
- Staying in care will help you/your child stay healthy
- Get regular heart checks even if you/your child feels well
  - New heart problems can happen with no symptoms
  - Early treatments can preserve health
- Most people with (condition) do well but some get (problem) over time.
- If you have these symptoms, seek care
- ***Repeat every time, say different ways***
- ***Less important: Name of diagnosis, specifics of anatomy/surgeries***

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P/F Group Education Strategies

- One-on-One conversations
- Educational materials – print, audio, video
- Education Meetings
- Peer Education – teach each other
- Contact with older patients
  - Models of thriving
  - Examples of challenges

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Beyond the Basics

- Long-term outcomes/Level of risk
- Who needs care, how often, from whom
- What kind of care
- Pregnancy – risks, care needs

Beyond Education

- Advocate for needed services
- Information resource for broader community

Care Guidelines can be an Essential Resource

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CHD and RHD Care Guidelines

- Produced by Professional Associations/Governments
- Often Region/Country- specific
- Goals
  - Guide specific clinical decisions
  - Promote high-quality care
- Describe major health risks – overall, condition-specific
- Recommend specific actions based on existing evidence
  - More evidence = stronger recommendation
- Controversial - "Not enough evidence"; "I do it differently", "That won't work here"

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How P/F Organizations can use

- **Organizational/Leader Knowledge**
  - Summary of existing research (Epidemiology, Health risks)
  - High-level recommendations
    - Recommended care schedule – who, how often, from whom
- **Guide Patient/Family Education**
  - "Translate" key information to patient-ese
- **Guide Advocacy**
  - Ask for needed services
- **Gives authority** – *"Experts recommend…"*
- **Aspirational –** what it should be like
- **Medical Advisors can identify/interpret**

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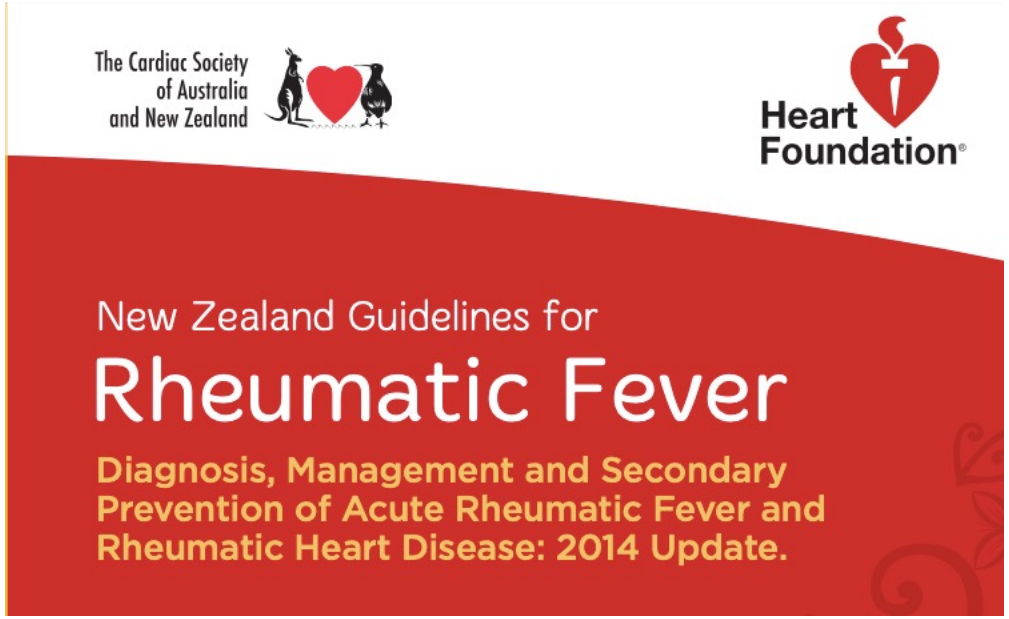

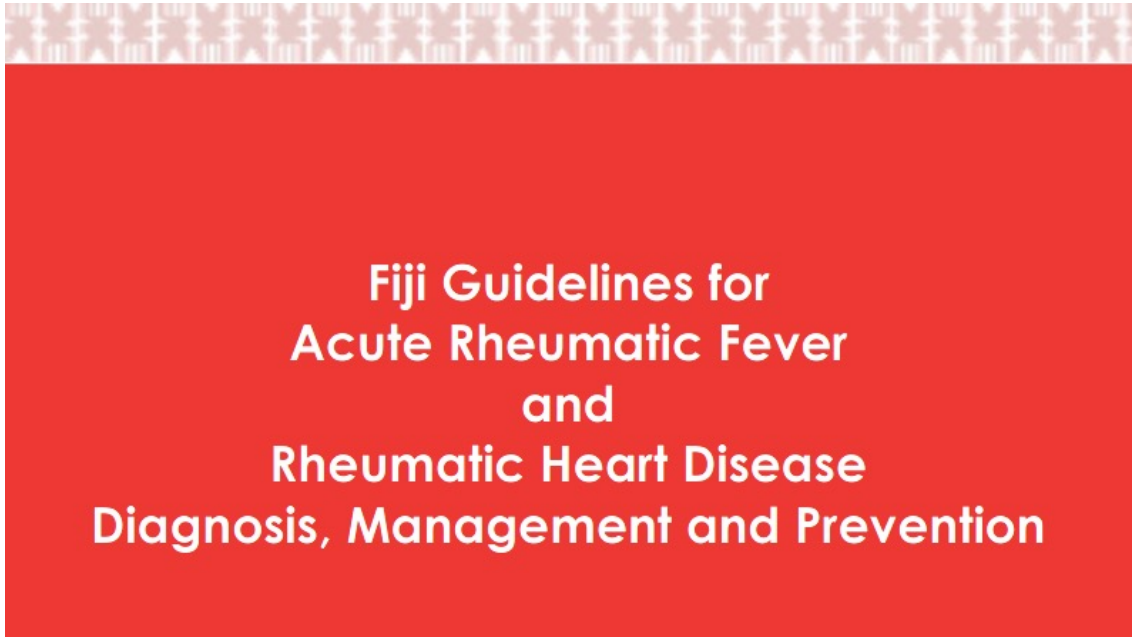

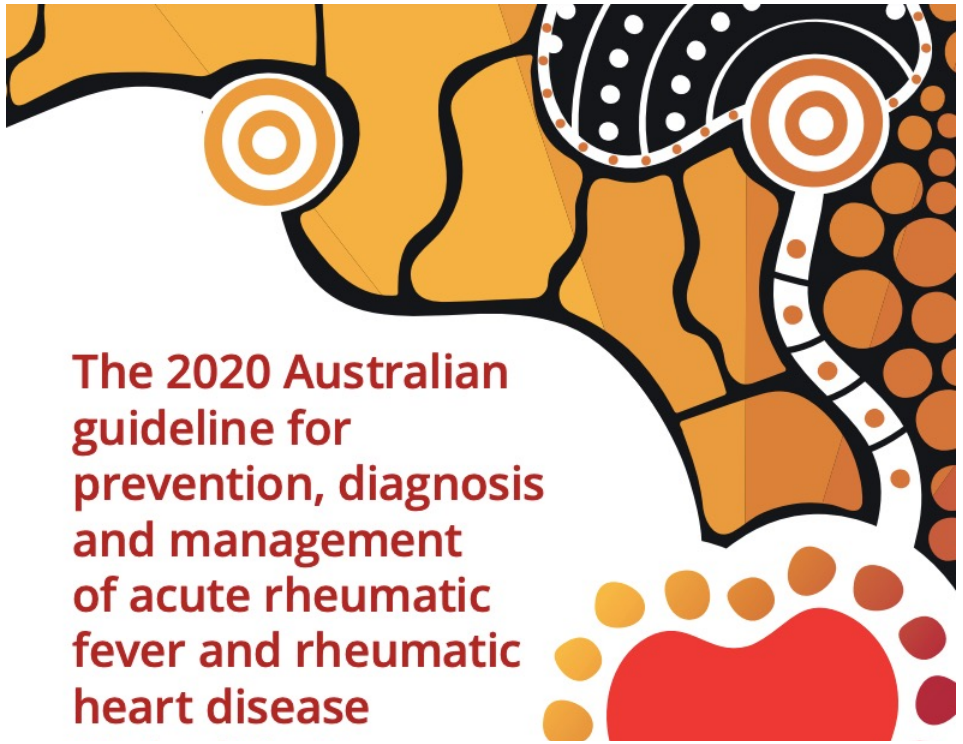

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AHA/ACC GUIDELINE

2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines

The Cardiac Society of Australia and New Zealand

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

ESC European Society of Cardiology
European Heart Journal (2021) **42**, 563–645
doi:10.1093/eurheartj/ehaa554

ESC GUIDELINES

2020 ESC Guidelines for the management of adult congenital heart disease

The Task Force for the management of adult congenital heart disease of the European Society of Cardiology (ESC)

- **All downloadable free**
- **Large-scale agreement**
- **Small differences**
  - **Specific treatments**
  - **Care intervals**

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Recommended Frequency of Care - RHD

DIAGNOSIS	RECOMMENDED FOLLOW-UP PLAN[†]
Priority 1	
Severe RHD[‡] High risk post-valve surgical patients[§] ≥ 3 episodes of ARF within the last 5 years Pregnant women with RHD (of any severity) may be considered Priority 1 for the duration of the pregnancy Children ≤ 5 years of age with ARF or RHD	Specialist review: at least 6 monthly Echocardiogram: at least 6 monthly Medical review: at least 6 monthly Pregnant: see *Figure 12.1 for care pathway* Dental review: within 3 months of diagnosis, then 6 monthly
Priority 2	
Moderate RHD[‡] Moderate risk post-valve surgical patients[§]	Specialist review: yearly Echocardiogram: yearly Medical review: 6 monthly Dental review: within 3 months of diagnosis, then 6 monthly
Priority 3	
Mild RHD[‡] ARF (probable or definite) without RHD, currently prescribed secondary prophylaxis Borderline RHD currently prescribed secondary prophylaxis Low risk post-valve surgical patients[§]	Specialist review: 1 – 3 yearly Echocardiogram: children ≤ 21 years: 1-2 yearly, > 21 years: 2-3 yearly Medical review: yearly Dental review: yearly

https://www.rhdaustralia.org.au/arf-rhd-guideline

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Recommended Frequency of Care - CHD

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Scoring System – Anatomy + Physiologic Stage

1. Simple Defects	2. Moderate Complexity	3. High Complexity
Small unrepaired ASD/VSD Mild pulmonic stenosis Repaired PDA – No additional problems Repaired ASD/VSD – No associated problems	Unrepaired Moderate/ Large ASD/PDA Repaired ASD/VSD/PDA – Associated problems AVSD Coarctation of the Aorta Tetralogy of Fallot Moderate/severe congenital valve disease Anomalous arterial/venous connections Congenital fistulas	Double-outlet Ventricle Single Ventricle Atresia – all forms Fontan Transposition disorders Interrupted aortic arch Truncus Arteriosus Other abnormalities of ventricular/arterial connection

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Physiological Stages

A	B	C	D
NYHA Stage I No hemodynamic or anatomic sequelae No arrhythmia Normal Exercise Function Normal liver/kidney/lungs	NYHA Stage II Mild hemodynamic sequelae Mild valve disease Trivial shunt Arrhythmia not needing treatment Cardiac limitations to exercise	NYHA Stage III Significant valve disease Moderate ventricular dysfunction Moderate aortic enlargement Venous/Arterial stenosis Hypoxia/Cyanosis Significant shunt Arrhythmias controlled with treatment Pulmonary hypertension Liver/kidney dysfunction controlled with treatment	NYHA Stage IV Severe aortic enlargement Uncontrolled arrhythmia Severe hypoxemia Severe Pulmonary hypertension Eisenmenger Syndrome Uncontrolled liver/kidney dysfunction

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- 1-A needs ACHD care every 3-5 years
- All others between 2 years and 3 months

Table 31. CCTGA: Routine Follow-Up and Testing Intervals (Table view)

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	12	12	6–12	3–6
ECG	12	12	12	12
TTE†	12–24	12	12	12
Pulse oximetry	As needed	As needed	Each visit	Each visit
Holter monitor	12–60	12–60	12–36	12
CMR‡/CCT§	36–60	36–60	12–24	12
Exercise test‖	36–60	36–60	12–24	12

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Pregnancy in RHD and CHD

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- Many people with RHD and CHD can have children safely **IF they get needed special care**
- People with severe heart failure, PH, cyanosis – pregnancy very high risk/discouraged
- **Pre-pregnancy consult** should be done
  - Correct problems before pregnancy
- Women with moderate/ severe CHD/RHD **need on-going special monitoring**
- Baby should be screened for CHD

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Thank You!

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