



## Heart Failure in Adult Congenital Heart Disease

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## Overview of Congenital Heart Disease

- CHD is the #1 birth defect, affecting one out of 120 babies born
- More than 85% of children undergoing CHD surgery will now survive to adulthood
- Ratio of adults: children with CHD is now 2:1



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## Overview of Congenital Heart Disease

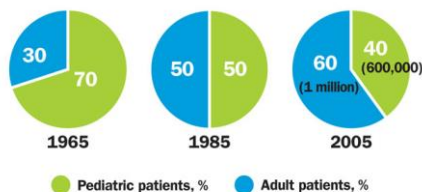
- Numbers are expected to keep growing by about 5% per year
- Most congenital heart disease is not cured by surgery
- Most children with "repaired" CHD are well-many important complication only arise in adulthood



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## Burden of Adult CHD

### Changing Proportion of Pediatric & Adult CHD

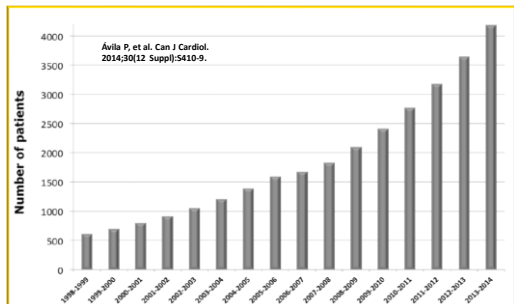


Adapted from: Williams RG, et al. Report of the National Heart, Lung, and Blood Institute Working Group on Research in Adult Congenital Heart Disease. J Am Coll Cardiol. 2006;47(4):703-707.



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## Patients followed at the Montreal Heart Centre 1998-2014



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## ACHD Prevalence by DHB

- Auckland DHBs: 6600
- Canterbury: 2200
- Bay of Plenty: 950
- Taranaki: 500



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So what does the evidence tell us?

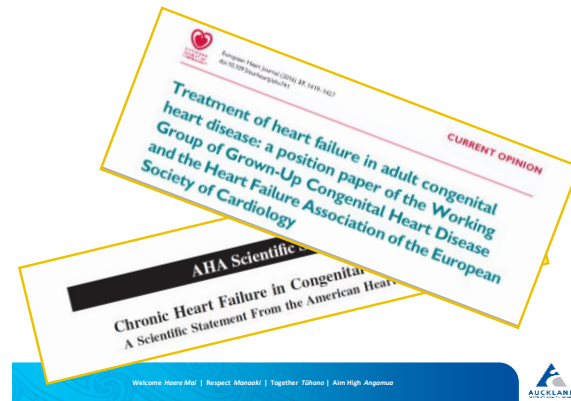
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So what does the evidence tell us about HF in the adult congenital population?

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Heart failure in ACHD

- More cumulative survival, means more pts at risk of HF
- HF leading cause of death in adults with CHD
- Pts admitted with HF have a five fold risk of death
- Significant mortality and hospitalisations

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Mechanisms of HF in CHD

- Volume overload
- Pressure overload
- Ventricular failure related to intrinsic myocardial dysfunction
- Pulmonary hypertension

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## Mechanisms of HF in CHD

- Systemic arterial hypertension
- CAD
- Cyanosis
- Intractable atrial arrhythmias

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## Signs and symptoms of HF

Symptoms of systemic ventricular failure	Signs of systemic ventricular failure
Fatigue	Third or fourth heart sound (gallop)
Breathlessness	Laterally displaced apical impulse
Dry cough especially lying flat	Pulmonary crepitations
<b>Often subtle signs such as decreasing weight, cachexia</b>	
Weight gain (>2kg/week)	Ascites
Loss of appetite	Pitting leg oedema, sacral oedema, scrotal oedema
Reduced exercise tolerance	
Increased abdominal girth	
Symptoms of congestive (biventricular) failure	Signs of congestive (biventricular) failure
Combined systemic and sub-pulmonary symptoms	Combined systemic and sub-pulmonary signs

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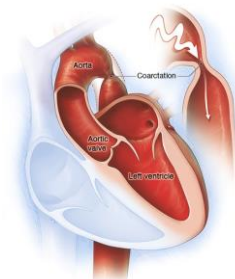
## Recommendations

- Anatomic and haemodynamic evaluation
- Look for reversible or repairable structural abnormalities
- Cardiopulmonary exercise testing
- Look for other causes e.g. anaemia, PHTN

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## Coarctation of the Aorta

- Recurrent coarctation
- Aneurysm
- Hypertension

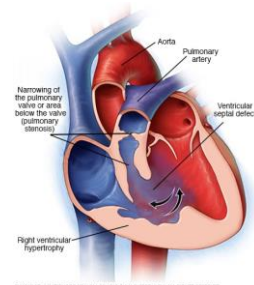


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## Tetralogy of Fallot

- RV dilatation and dysfunction
- Arrhythmias
- Impaired functional capacity



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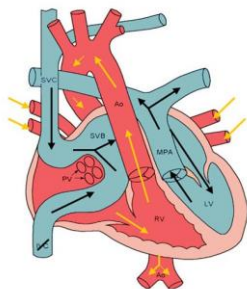
## Pulmonary valve replacement

- Right ventricular indications
- RV diastolic volume index >150-170ml/m<sup>2</sup>
- RV systolic volume index >80ml/m<sup>2</sup>
- RVEF<40%

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## Mustard Senning -TGA Auckland District Health Board

- ❑ RV dysfunction
- ❑ Tricuspid regurg
- ❑ Sinus node dysfunction
- ❑ Tachyarrhythmias
- ❑ Baffle complications



## RV function after Mustard/ Senning Auckland District Health Board

**Table 2** Diagnostic tests

	1990 <sup>10</sup>	2001 <sup>7</sup>	2012	P-value <sup>8</sup>	
Echocardiogram	n = 58	n = 53	n = 47		
RV systolic function normal	40 (69%)	3 (6%)	1 (2%)	<0.001	0.3

Cuypers et al. (Erasmus, Netherlands)  
*Eur Heart Journal* 2014; 35(25); 1666-

## Can we extrapolate standard HF therapy to CHD pts? Auckland District Health Board

- ❑ Lesion specific
- ❑ Data sparse
- ❑ Inconclusive
- ❑ Poor end points

