

# Heart Failure in Patients with Congenital Heart Disease (CHD)



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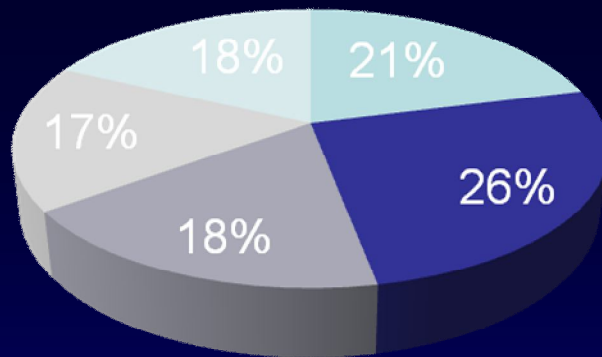


# Introduction

- HF is common in older patient populations
- HF is becoming more common in ACHD
- With advancing age patients with CHD develop ventricular dysfunction

# Cause of Death in ACHD

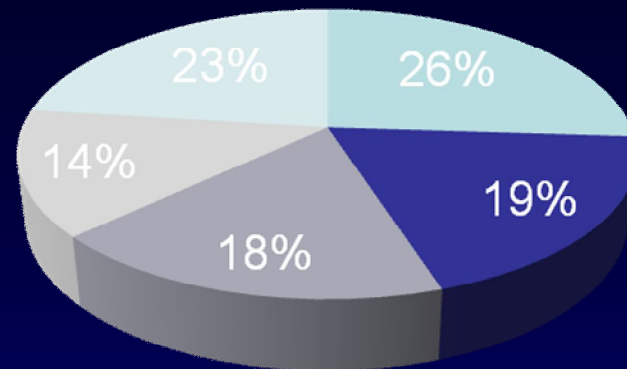
- CHF 21%
- SD 26%
- Peri-op 18%
- Non-cardiac 17%
- Other CVS 18%



N = 2609 pts  
 17 year data collection (1981 to 1998)  
 Nber of decease: 199 (7.6%)

Oechslin AJC 2000

- CHF 26%
- SD 19%
- Cardiac various 18%
- Vascular 14%
- Non cardiac 23%



N= 6933  
 Data collection 2002 to 2008  
 Nber of decease: 197 (2.8%)

Verheugt et al, EHJ 2010

# ACHD Developing HF (1)

	Description	Anatomy	Number Studied	Age at Assessment	Percent Symptomatic
Graham et al 2000	Retrospective analysis of record	CCTGA	182	45	67%
Piran et al 2002	Consecutive & prospective study	Mustard CCTGA Fontan	90 65 33	15 y post surg	22% 32% 40%

## ACHD Developing HF (2)

	Description	Anatomy	Nber	Age at Assessment	echo	Percent Symptomatic
Ross-Hellekin et al 2004	Prospective study	TGA-Mustard	91	15y post surg.  25y post surg.	69% normal  6% normal	75 % NYHA class 1  24% NYHA class 1

## Other Potential Lesions Presenting with HF

### Presenting with left HF symptoms

- Mitral valve disease (MR)/regurgitant LAV
- Aortic valve disease (AR)
- IHD
- Coartation & hx of hypertension

### Presenting with right HF symptoms

- Large ASD – elderly
- Tricuspid valve disease (e.g. Ebstein's)
- Pulmonary valve disease (PR, ventriculotomies, transannular patch )
- RV to PA conduit stenosis (e.g. Rastelli)

# Mechanisms of HF

- Unlike IHD, causing LV systolic dysfunction because of sudden myocyte loss, HF in the ACHD is caused by multiple mechanisms:
  - Long standing pressure and /or volume overload due to obstructive or regurgitant lesions, shunting.
  - Recurring arrhythmias
  - Cardiopulmonary bypass (previous era myocardial protection suboptimal)
  - Ischaemia (e.g. Injury during surgical repair)

# Evidence-Based Medical Therapy in HF

- Published consensus guidelines on the management of ACHD do not address the issue of medical therapy for HF
- Limited literature available on the benefits of pharmacological therapy in congenital HF



# Medication Trials (1 of 2)

	Pro/ retro	Agents	TGA CCTGA	No	Mean Age	Follow- up (Months)	Results
<b>ACEi</b> Hetchter et al, 2001	Retro No control	Various ACEi	TGA	14	31	6	No significant changes; Exercise time improvement in some pts
Robinson et al, 2002	Pro	Enalapril	TGA	6	13.8	12	No significant changes
<b>ARB</b> Lester et al, 2005	Pro	Losartan	TGA	7	>13	2	Improvement of exercise time & systemic AV valve regurgitation
Dore et al, 2005	Pro	Losartan	TGA & CCTGA	29	30.3	3.2	No significant changes
Van der Bon et al, 2013	Pro. multi centre double blind placebo controlled trial	Valsarta n	TGA & CCTGA	88	33	36	No significant changes

Pro, prospective study design; Retro, retrospective study design; AV valve, atrio-ventricular valve

# Medication Trials (2 of 2)

	Pro/ retro	Agents	TGA CCTGA	No	Mean Age	Follow-up (Months)	Results
<b>BB</b> Doughan et al, 2007	Retro	Carvedilol & Metoprolol	TGA	31	29	4	Improvement in NYHA class
Giardini et al, 2007	Retro	Carvedilol	TGA & CCTGA	8	26	12	Improvement of RV function & exercise tolerance

# Cardiac Resynchronization Therapy

- Little data
- 4-9% patients with systemic RV are potentially eligible for CRT (Diller et al, 2006)
- Small studies suggest that CRT may be beneficial (Janousek et al, 2001 & 2004)

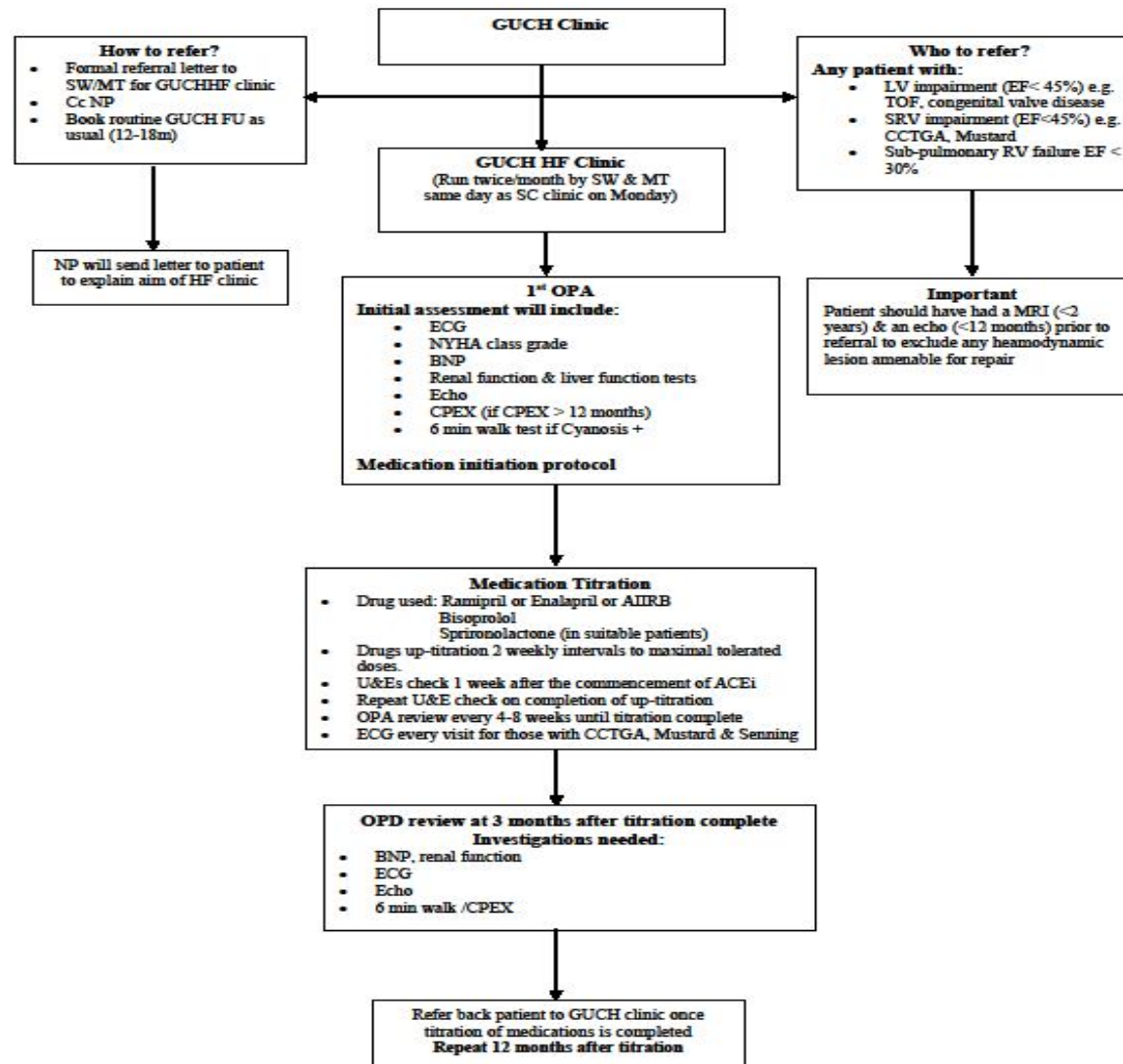
# Heart Transplant

- Very small percentage of heart transplants are undertaken on ACHD in UK
- Transplantation is often more complex and challenging (higher early mortality, high level of antibodies, technically more difficult, pulmonary hypertension...)

# Aim of HF Service for ACHD

- Assess severity of ventricle function
- Optimise HF medical therapy & assess for potential device therapies
- Refer patients for heart transplant
- Respond to psycho-social needs
- Introduce EOL issues and refer to PC team

## Management of Patients with Grown-Up Congenital Heart Disease and Heart Failure



# Role of the CNS in HF (1 of 4)

## To provide education

- The basic principles of caring for patients with HF can be applied to both young and old patients.
- Keys elements that patients with HF needs to be educated in:
  - Condition.
  - Treatment regimens – Medications and potential side- effects, treatment compliance, fluid restriction, diet, exercise.
  - Self-monitoring – daily weight monitoring, detection of early signs and symptoms of decompensation

# Role of the CNS in HF (2 of 4)

## To adjust and optimise HF drugs

- To monitor patients through regular telephone contact and re-adjust drugs when patients present early symptoms of deterioration.
- To optimise HF drugs through locally agreed protocols – (done in clinic or over the telephone).



## Drug Titration Schedule for Heart Hospital GUCH HF Clinic

Surname: XXXXX  
 First Name: XXXX  
 Hospital Number: XXXXX  
 Date of Birth: XX/XX/XX  
 Diagnosis: TOF  
 Pacemaker/AICD: AICD

Symptom change that must be documented:  
 - breathlessness  
 -fatigue  
 -postural dizziness  
 - cough or wheeze

Date	Symptom change	Ankle oedema	Wgt (kg)	BP	HR	NA	K	Ur	Creat	Safe titration	Drug	Dose	Next visit
18.06.07			73.9	116/85	77	144	3.7	3.5	64	-	Ramipril introduced	1.25mg Od	09..07.07
09.07.07	Dry cough	No	74	112/85	72	-	-	-	-	-	Ramipril changed to Candesartan	4mg od	06.08.07
06.08.07	No	No	73.3	110/80	60	140	4.1	3.6	53	Yes	Candesartan increased	8mg od	15.09.07
19.08.07	No	No	70.9	102/67	73	-	-	-	-	Yes	Candesartan increased	12mg od	17.12.07
02.09.07													

Y=Yes  
 N=No

Any questions related to drug titration should be directed to Nathalie Picaut, GUCH clinical nurse specialist

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Address: GUCH Unit, The Heart Hospital, ~~05.05.09~~ 16-18 Westmoreland Street, London W1G 8PH

# Role of the CNS in HF (3 of 4)

## To respond to psychological needs

- To diagnose anxiety and depression which is common in patients with HF due to loss of independence, self-esteem
- To provide support & counseling to patients
- To refer patients to counselor or psychologist when needed

# Role of the CNS in HF (4 of 5)

## To respond to social needs

- Key elements that may need to be addressed
  - Employability
  - Benefits
  - Insurability
  - Home care

# Role of the CNS in HF (5 of 5)

## To initiate End-of-life discussions and refer to Palliative Care Service

- End-of-life discussions can be challenging when dealing with young patients.
- Potential barriers to end-of-life communication include:
  - Difficulties to predict outcome.
  - Difficulties to recognize and admit that there is no further intervention that can save patient.
  - Young patients are comfortable with medical technologies and expect options that will save their life.
  - Young patients have not been exposed to death & grief.
  - Young patients are not expecting to die young

# Summary

- Increasing number of ACHD develop ventricular dysfunction
- HF services must be expanded, evaluated & results published
- Research is needed to develop local & national guidelines for the PC approach in ACHD
- ACHD nurse specialists can play a pivotal role in improving the care and providing evidence based to manage patients with CHD and HF