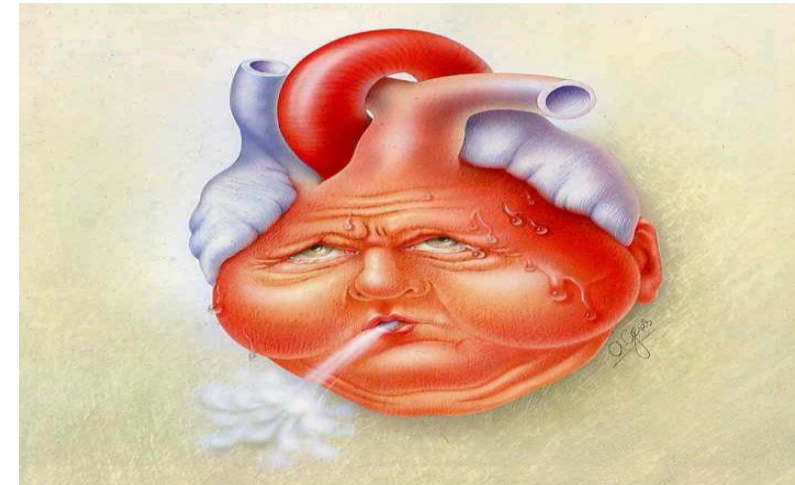


Heart Failure in Adult Congenital Heart Disease

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June 2020

Aims

- To gain an enhanced understanding of heart failure in the context of congenital heart disease
- To increase awareness of how heart failure affect adult congenital heart disease



Objectives

- Review the evidence available regarding heart failure in the congenital heart disease
- Provide practical advice and self care strategies
- Discuss the stages in treatment

Prevalence of Heart failure in CHD

- Many congenital heart disease patients, often with increased complex category of congenital heart disease have a higher chance of developing a heart failure.
- Many congenital heart patients on medications live very full and active lives.

Definition

“In physiological terms, heart failure is a syndrome characterised by either or both pulmonary and systemic venous congestion and / or inadequate peripheral oxygen delivery, at rest or during stress, caused by cardiac dysfunction”.

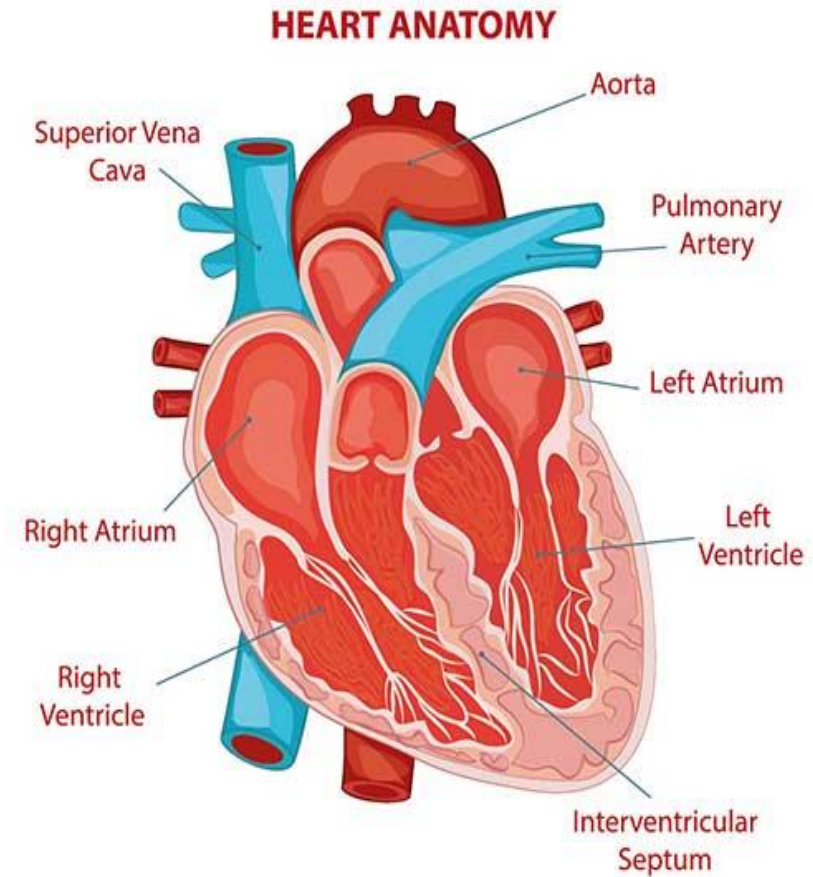
American Heart Association - 2016

Heart Failure (H F)

- Can happen at any time
- Common cause of death in ACHD patients - more common is systemic Right Ventricular (RV) failure.

Common Congenital Heart Defects

- Atrial septal defect
- Ventricular septal defect
- Tetralogy of Fallot
- Transposition of the great arteries
- Coarctation of the aorta
- Aortic stenosis
- Pulmonary stenosis



Complex Lesions

- Fontan operation
- Mustards procedure
- Single ventricle
- Hypoplastic Left Heart
- Pulmonary atresia
- Double outlet right ventricle (DORV)
- Double outlet left ventricle (DOLV)
- Transposition of the great arteries

NURSING MNEMONICS & TIPS

RIGHT-SIDED HEART FAILURE "AW HEAD"

A

ANOREXIA & NAUSEA

Results from the venous engorgement and venous stasis within the abdominal organs.



W

WEIGHT GAIN

Due to retention of fluid.



H

HEPATOMEGALY

Results from the venous engorgement of the liver; increased pressure may interfere with the liver's ability to function.



E

EDEMA (BIPEDAL)

Pink or blood-tinged sputum may be produced.



A

ASCITES

Is the accumulation of fluid in the peritoneal cavity; increased pressure within the portal vessels forces fluid into the abdominal cavity.



D

DISTENDED NECK VEIN

Increased venous pressure leads to distended neck veins.

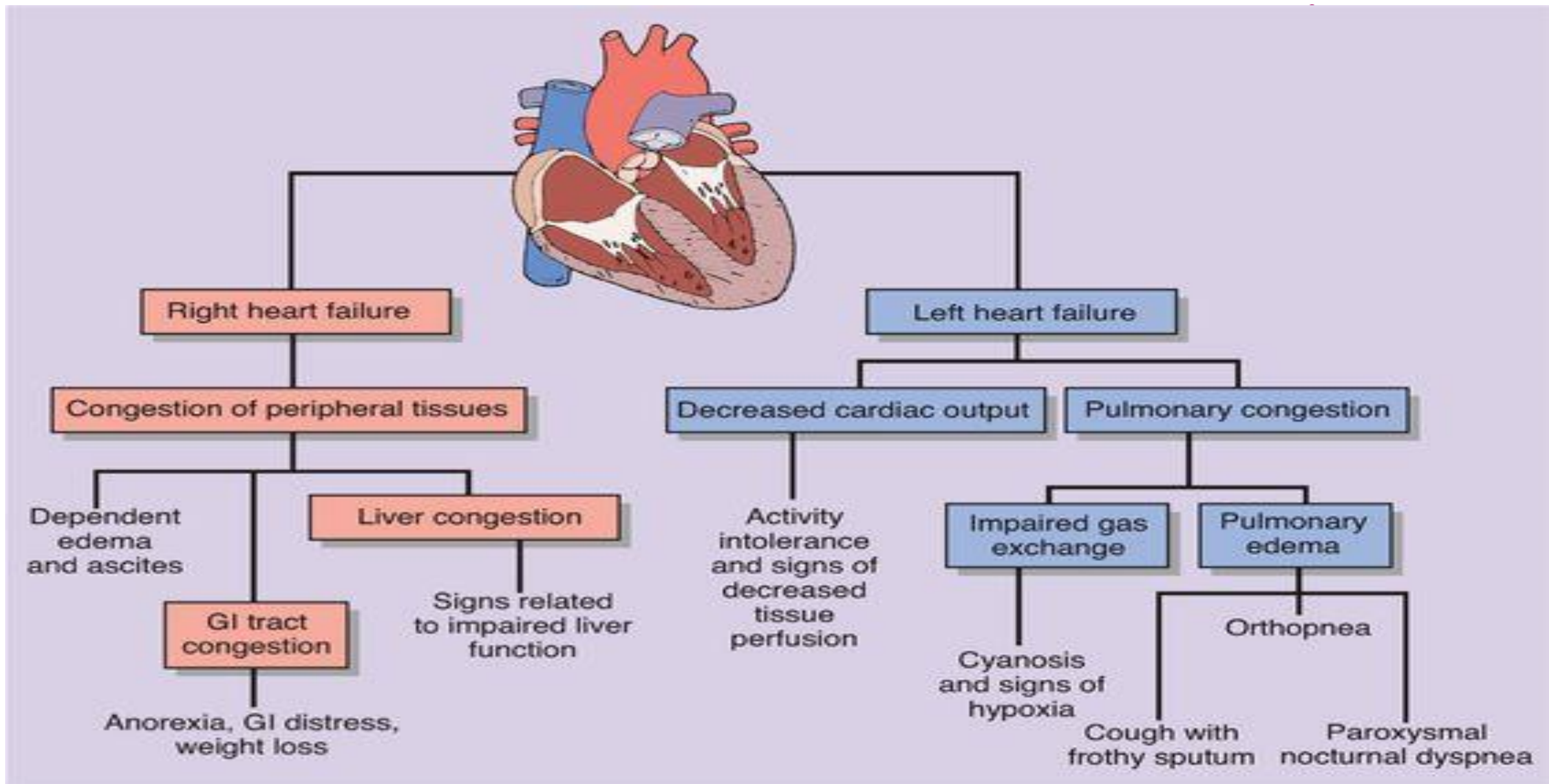


LEARN MORE: RIGHT-SIDED HEART FAILURE

When the right ventricle fails in right-sided heart failure, congestion in the peripheral tissues and the viscera predominates. This occurs because the right side of the heart cannot eject blood and cannot accommodate all the blood that normally returns to it from the venous circulation. Right-sided heart failure primarily produces systemic signs and symptoms.

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<https://www.heart.org/en/health-topics/heart-failure/what-is-heart-failure/types-of-heart-failure>

CONGENITAL HEART DISEASE

AT RISK OF HEART FAILURE

HEART FAILURE

Stage A

At high risk of HF but without structural heart disease or symptoms of HF

Structural Heart Disease

Therapy

- Early identification of predisposing factors
- Risk reduction

Stage B

Structural heart disease but without signs of HF

SYMPTOMS OF HEART FAILURE

Therapy

- As in Stage A

Selected patients:

- ACEi
- ARB
- BB
- AICD

Stage C

Structural heart disease with prior or current symptoms of HF

Refractory HF symptoms at rest

Therapy

- As in Stages A and B

Routine use:

- Diuretics if Fluid retention
- ACEi
- BB

Selected patients:

- Aldosterone antagonist
- ARBs
- Digitalis
- Hydralazine/nitrates
- CRT-P
- AICD

Stage D

Refractory HF requiring specialized interventions

Therapy

- Appropriate measures from Stages A, B and C
- Decide appropriate level of care

Options:

- Compassionate end-of-life care/ hospice
- Extraordinary measures:
 - Heart transplant
 - Chronic inotropes
 - Permanent mechanical support
 - Experimental surgery or drugs

Congenital heart disease:

- Surgical/ Percutaneous repair of hemodynamic lesions
- Iron supplementation
- Exercise training

CHD-related Pulmonary Hypertension

- Targeted Pulmonary Hypertension Therapies (ERAs and/or PDE-5i and/or Prostanoids)

1st line treatment

Medication

- Diuretics
- Beta-Blockers
- Angiotensin Converting enzyme inhibitors (ACE)
- Angiotensin receptor blockers (ARB)
- Mineralocorticoid receptor antagonist (Spironolactone)
- Digoxin
- *Ivabradine (Patients need to be in sinus rhythm).
- *Hydralazine / nitrates (EOL care).

2nd line treatment

- Biventricular Pacemakers
- Cardiac resynchronization therapy (CRT) CRT- P / CRT - D
- Implantable cardioverter defibrillator (ICD)



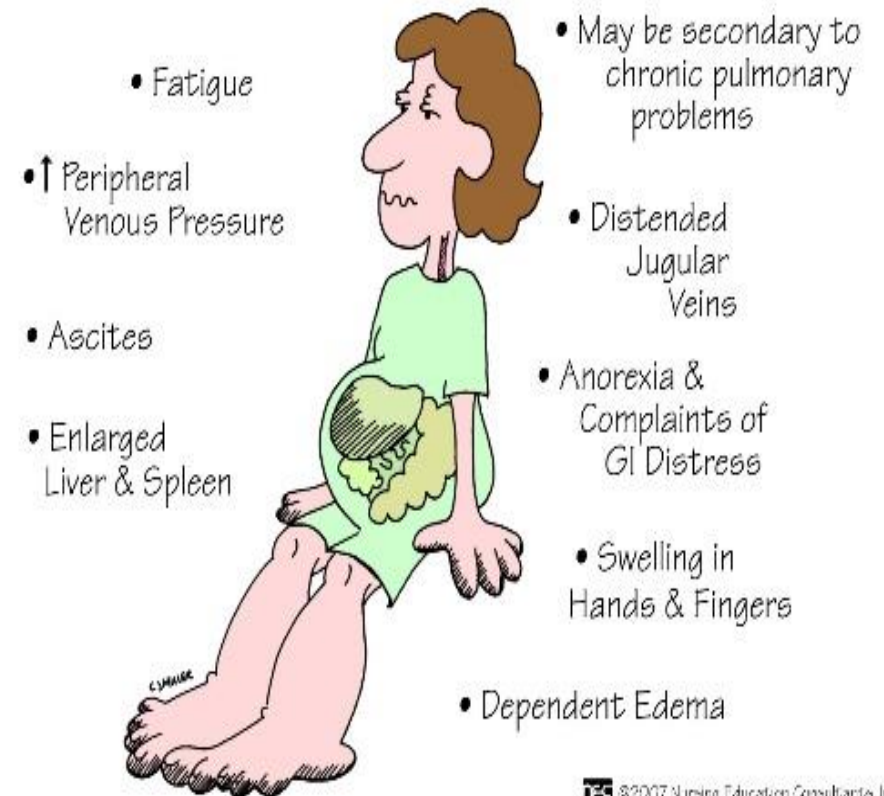
Self care strategies

Education is Key

- Daily Weight
- Is there a need for fluid restriction?
- Diet – low salt food
- Take medication as prescribed daily
- Check for swelling – Feet, ankles, legs, hands and stomach.
- Hepatomegaly

RIGHT SIDED ♥ FAILURE

(Cor Pulmonale)



BNP – NT-pro BNP

- Natriuretic peptides = Hormones that are made and released by the heart
- Known to rise in Heart Failure
- Higher the level the more severe the heart failure
- Raised BNP in pulmonary hypertension may be misinterpreted as representing left heart failure but ^ dyspnea can be due to pulmonary systemic disease
- Levels reduced in overweight patients
- With certain medication

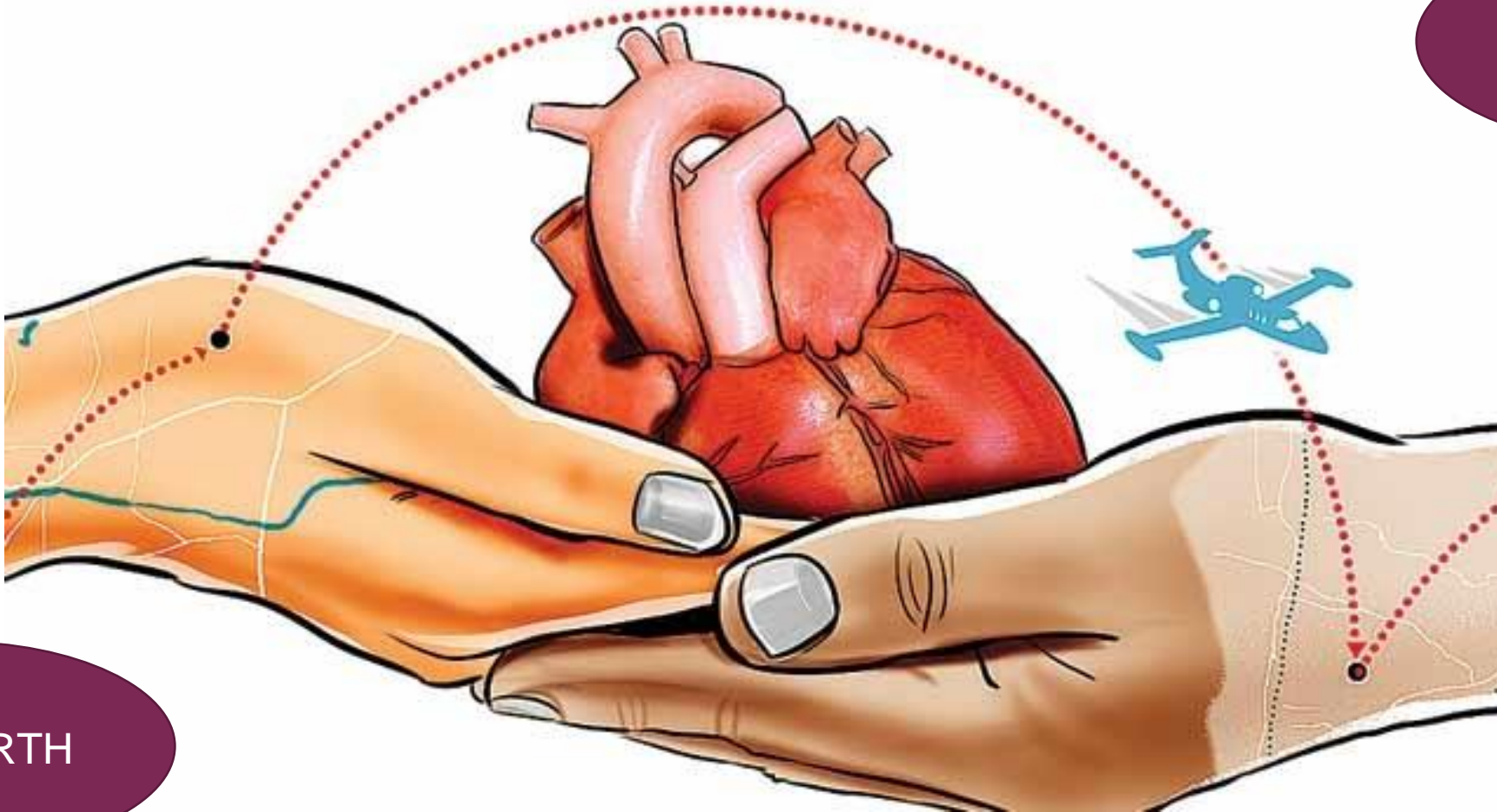
** Not widely used in ACHD – Aware of the etiology of the heart – determining the cause is important**

3rd line treatment – Heart Transplant



Transplant referral

NEWCASTLE



PAPWORTH

Parallel planning

Parallel planning is fundamental in palliative care. Essentially,

It is:

- Hoping for the best, but planning for the worse.
- Planning for living and planning for dying. In the same manner that you are equally planning for different circumstances.

Palliative and End of Life Care in Heart Failure



12 Palliative Care and Bereavement

- **sets out:** how to provide support at end of life and how to manage communication with families around the end of life
- **Challenge** : difficult conversations, patient, parents spouse, family and children
- Intense telephone advice
- End of life pathway
- Palliative care teams
- GP support
- **Network palliative care toolkit**



What is palliative care ?...

- Palliative care is the active, total care of patients whose disease is not responsive to curative treatment. Palliative care takes a holistic approach, addressing physical, psychosocial and spiritual care, including the treatment of pain and other symptoms. Palliative care is interdisciplinary in its approach and encompasses the care of the patient and their family and should be available in any location including hospital, hospice and community.
- Palliative care affirms life and regards dying as a normal process; it neither hastens nor postpones death and sets out to preserve the best possible quality of life until death.

Network Palliative Care Toolkit

Clinical Guideline

PLANNING PALLIATIVE & END-OF-LIFE CARE FOR CARDIAC PATIENTS ACROSS THE CARDIAC NETWORK TOOLKIT

SETTING Congenital Heart Disease Network for South Wales and South West

FOR STAFF All cardiologists, surgeons, junior doctors, nursing staff and allied health care professionals

PATIENTS All Congenital Heart Disease patients within the CHD Network regardless of location of care

EXECUTIVE SUMMARY:

This toolkit has been written in response to the Congenital Heart Disease (CHD) Standards and Specifications, published by NHS England in 2016. The standards consider the lifespan care of patients born with CHD and seek to ensure high quality, equitable and consistent care for all patients across the Network in South Wales and the South West. The standards are divided into several sections and section L covers palliative care. The standards cover the Level 1 specialist surgical centre (Bristol), Level 2 specialist cardiology centre (Cardiff) and the many level 3 centres across the network.

- Can be found on SWSWCHD Network website under professional resources
- www.swswchd.co.uk

<http://chdn.webboxdemo.co.uk/>

Welcome to the Congenital Heart Disease Network South Wales and South West

We proudly support over 6,500 children and 8,000 adults with a congenital heart condition.

[Read More >](#)

Babies and Children

When your baby or child is diagnosed with a cardiac condition you may not know what to

Teenagers/Young People

When you are diagnosed with a cardiac

Adults

When you are diagnosed with a cardiac condition you may not know what to expect.

References:

- ESC Guidelines -

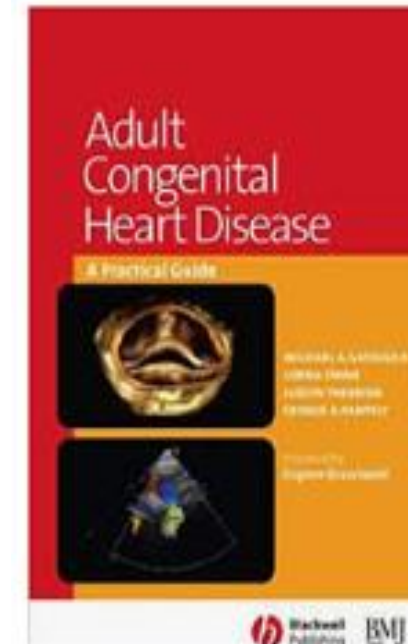
<http://eurheartj.oxfordjournals.org/content/31/23/2915.full.pdf>

- American guidelines -

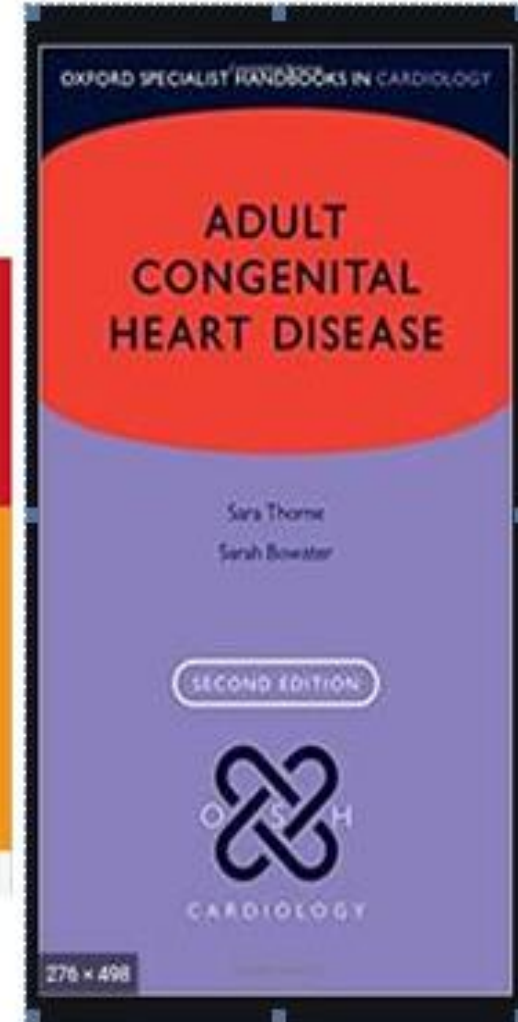
<https://www.ahajournals.org/doi/epub/10.1161/CIR.0000000000000603>

- Network Palliative Tool kit

<https://www.swswchd.co.uk/en/page/clinical-information-palliative-care>



Adult Congenital Heart...
amazon.co.uk



<https://www.heart.org/en/health-topics/heart-failure/what-is-heart-failure/types-of-heart-failure>

Miyata, Masaaki; Kihara, Takashi; Kubozono, Takuro; Ikeda, Yoshiyuki; Shinsato. *Journal of cardiology*; (2008) vol. 52 (no. 2); p. 79-85



Thank you