

# Adult Congenital Heart Disease Interactive Case Studies



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CNS

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# Aims of the talk:

- To gain new knowledge and insight into the health needs of the complex congenital heart disease patient with Fontan circulation and the reality faced by this patient population group
- Develop an awareness of the care needs for these complex congenital heart disease patients

# Areas for discussion:

- Different types of Fontan procedures
- Mortality age compared with mortality in age- matched UK population of congenital heart disease patients
- How much have you learnt today? - Quiz time!

- ◆ Baffle: artificial wall created to route blood flow in a desired direction
- ◆ Balancing circulations: the effects of pulmonary and systemic blood flow ratio (referred to as the  $Q_p/Q_s$  ratio)
- ◆ Conduit: tube or artificial passage placed to carry blood from one area to another
- ◆ Fenestration: a small window or hole in the patch used to create the intra-atrial tunnel in a Fontan operation to help control elevated systemic venous pressure postoperatively
- ◆ Fontan circulation: diverting of venous blood from the right atrium to the pulmonary arteries, allowing blood flow to the lungs without it being pumped by the heart
- ◆ Isthmus ablation: a catheter-based procedure performed in the electrophysiology lab to eliminate conduction of atrial arrhythmias by using radiofrequency ablation to the cardiac isthmus in the right atrium
- ◆ Maze procedure: an open heart surgery performed to treat atrial fibrillation, whereby a number of incisions are made on the left and right atrium to form scar tissue that then disrupts the path of abnormal electrical impulses and prevents them from reoccurring
- ◆ Shunt: surgical creation of a new route for blood flow.

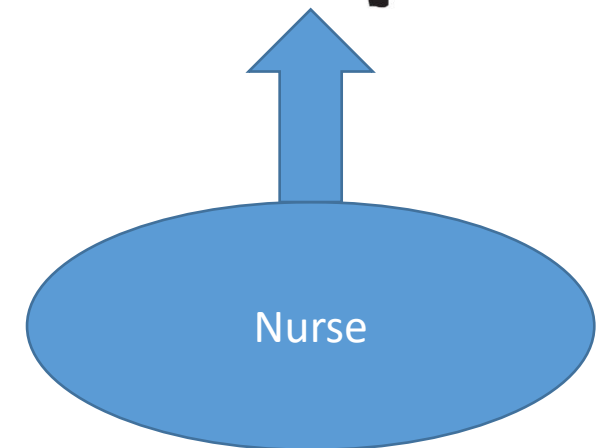


# Preconceptions:

There is a preconception that complex ACHD patient are scary to look after!....

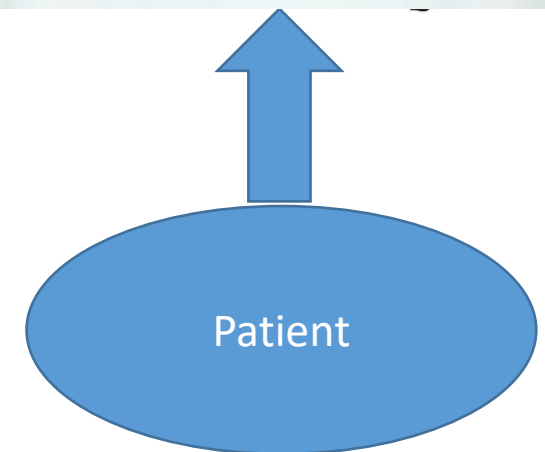
This is not  
true!....

AS LONG AS YOU TAKE TIME TO KNOW YOUR PATIENTS  
HISTORY AND ANATOMY!



# How the patient feels...

- Have been in and out of hospital many times through their life and would have had many operations as a child.
- Build up a good relationship – need to gain trust.
- Good clear communication. Open and honest



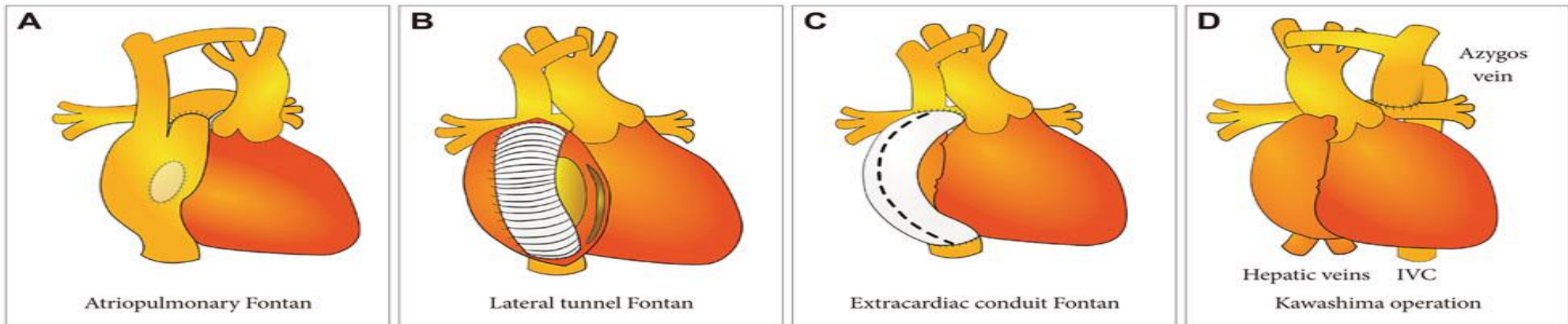
# Choussat's ten commandments:

Choussat et al. (1977) delineated selection criteria to define an ideal candidate for a Fontan procedure. They described the 10 following criteria, which are occasionally and facetiously referred to as the Ten Commandments for an ideal Fontan operation result.

- Age >4 years
- Sinus rhythm
- Normal systemic venous return
- Normal right atrial volume
- Mean pulmonary artery pressure <15 mm Hg
- Pulmonary arteriolar resistance <4 Wood units/m<sup>2</sup>
- Pulmonary artery–aorta ratio >0.75
- Left-ventricular ejection fraction >0.60
- Competent mitral valve
- Absence of pulmonary artery distortion

# Different Fontan techniques:

- Ventricularization of the Right Atrium (The original **Fontan's** Technique)
- Intracardiac total cavopulmonary connection (lateral tunnel)
- Extracardiac total cavopulmonary connection (extracardiac tunnel)
- Atriopulmonary connection (the original Kreutzer's Technique)







Remember:

Fontan operation is “non curable” therefore palliative operation...

Single ventricle:

- Hypoplastic Left Heart
- Pulmonary Atresia/Intact Ventricular Septum
- Tricuspid Atresia
- Double outlet right ventricle (DORV)
- Double outlet left ventricle (DOLV)

# Complications of Fontan circulation

- Exercise intolerance
- Ventricular failure
- Right atrium dilatation
- Arrhythmia
- Systemic and hepatic venous hypertension
- Portal hypertension
- Coagulopathy
- Pulmonary arteriovenous malformation
- Lymphatic dysfunction (eg, ascites, edema, effusion, protein-losing enteropathy, and plastic bronchitis).

|                      | Patient's age (years) |    |    |    |    |    |    |    |    |    | Age difference: |
|----------------------|-----------------------|----|----|----|----|----|----|----|----|----|-----------------|
|                      | 20                    | 25 | 30 | 35 | 40 | 45 | 50 | 55 | 60 | 60 |                 |
| ASD                  | 25                    | 26 | 32 | 38 | 42 | 47 | 52 | 57 | 61 | 61 | <2              |
| Valvar disease       | 29                    | 31 | 36 | 40 | 45 | 49 | 54 | 59 | 63 | 63 | 2-5             |
| VSD                  | 28                    | 30 | 36 | 40 | 44 | 49 | 53 | 59 | 63 | 63 | 2-5             |
| Aortic Coarctation   | 32                    | 33 | 38 | 43 | 47 | 52 | 56 | 62 | 66 | 66 | 5-10            |
| AVSD                 | 33                    | 34 | 39 | 44 | 48 | 52 | 57 | 62 | 66 | 66 | 5-10            |
| Marfan syndrome      | 37                    | 38 | 42 | 46 | 50 | 54 | 59 | 64 | 68 | 68 | 5-10            |
| Tetralogy of Fallot  | 37                    | 38 | 42 | 47 | 50 | 54 | 60 | 65 | 69 | 69 | 5-10            |
| Ebstein anomaly      | 42                    | 43 | 47 | 51 | 54 | 59 | 63 | 68 | 72 | 72 | 10-20           |
| Systemic RV          | 46                    | 48 | 51 | 55 | 59 | 63 | 67 | 72 | 76 | 76 | 10-20           |
| Eisenmenger syndrome | 57                    | 58 | 62 | 65 | 69 | 73 | 77 | 81 | 84 | 84 | 20-30           |
| Complex CHD          | 58                    | 59 | 63 | 67 | 70 | 74 | 78 | 82 | 85 | 85 | 20-30           |
| Fontan               | 64                    | 65 | 68 | 72 | 75 | 78 | 82 | 86 | 91 | 91 | >40             |

# Cyanosis:



- Cyanosis results from an increase in circulating RBC as the body attempts to improve its oxygen carrying capacity.
- Increased viscosity can lead to thrombosis, stroke, embolus, infection brain abscess, PH, gout, gall stones, iron deficiency, arrhythmias, and renal dysfunction.
- Caution if NBM, IV fluids.

Evaluation of the failing or failed Fontan circulation requires knowledge of the anatomic substrate, surgical intervention details, cardiac imaging and assessment of haemodynamic status, assessment of rhythm status, and evaluation of other organ systems and metabolic function.

Deal, B. Jacobs, M. (2012).

# Red Flags and early warning signs of a failing Fontan:



- Exercise intolerance (> in ability to exercise ) or ^ NYH classification
- Cyanosis (Blue lips - decrease in SPO2)
- Liver complications - Ascites
- Arrhythmias



# Arrhythmia



- 32% of all patients post ACHD surgery have a sustained arrhythmia
- Important to note that arrhythmias in Fontan circulation are associated with serious complications including heart failure and death... Cardiovert back to regular rhythm ASAP

# Case Study 1

18 Y.O Seen in Y.P clinic for the 1<sup>st</sup> time –



Case Study: N. Bryant – 44 year old female to ensure patients confidentiality is maintained (NMC2009) his name has been changed.

Diagnosis:

- Double inlet left ventricle with pulmonary atresia and unrestricted ventricular septal defect
- Initial palliation with right BT shunt *1974*
- AP Fontan connection in *1987*
- Patch closure of tricuspid valve defect in 1990
- Attempted umbrella closure of right atrial to LV leak
- Atrial fibrillation
- Amiodarone induced thyrotoxicosis with subsequent radioactive iodine treatment and reintroduction of Amiodarone
- Symptomatic bradycardia - VVI pacemaker via coronary sinus

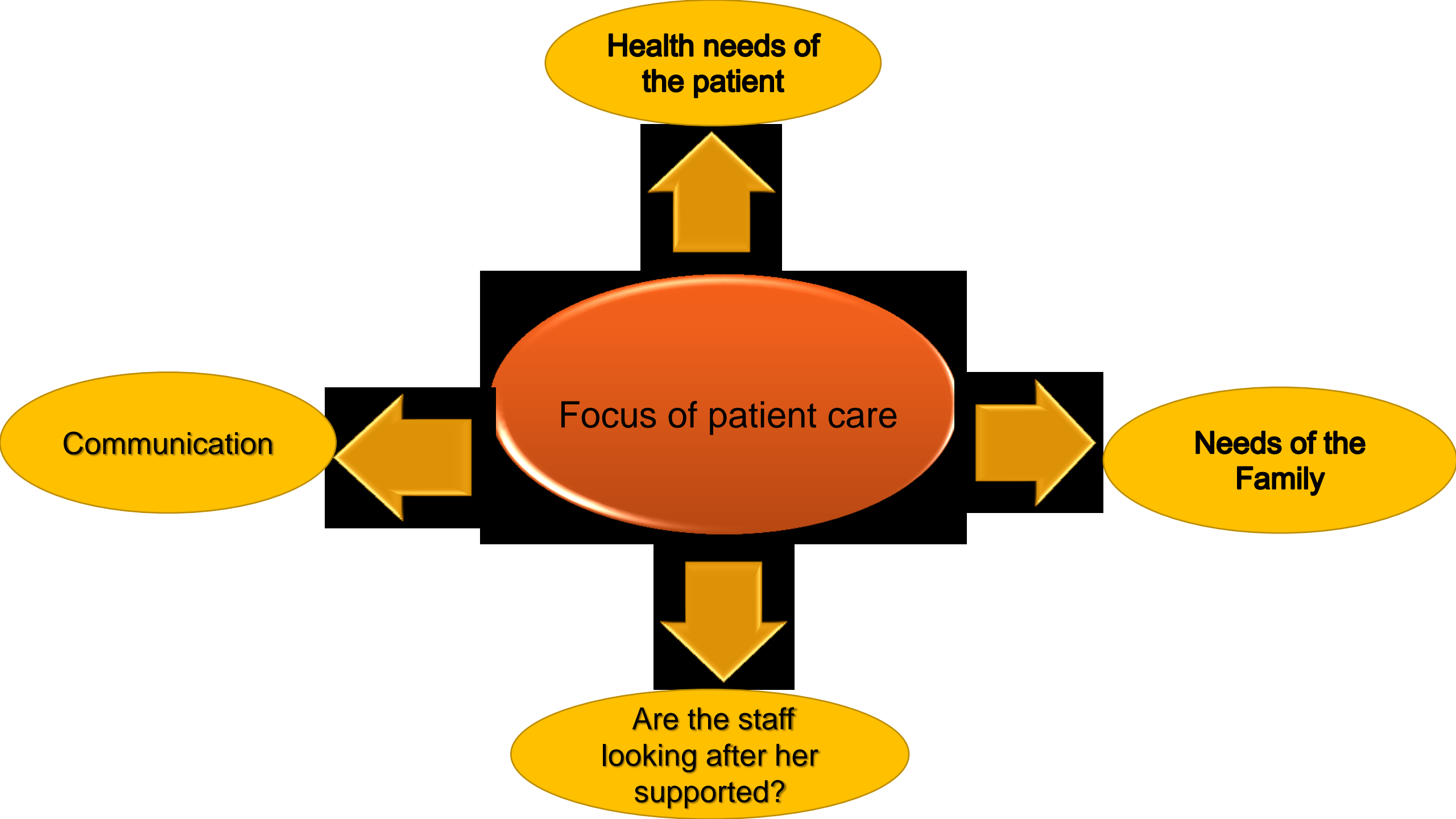
# comorbidities:

- Chronic venous ulcerations - legs
- Hypothyroidism
- Long-term organised laminated thrombus in right atrium with more recent roof position clot
- Recurrent vaginal bleeding - under gynaecologists

Patient called CNS advice line: Symptoms increasing ^

Admitted to hospital via A&E On clinical examination:

- Peripheral oedema to the thigh
- ECG:
- Echo: severe bi-ventricular impairment.



**Health needs of  
the patient**

**Focus of patient care**

**Needs of the  
Family**

**Are the staff  
looking after her  
supported?**

**Communication**

# Difficult conversations

- **Uncertainty** - Advanced care planning/ceiling of care. Support complex discharges.
- Ask “is is ok” to talk to them... “Is it a good time to talk”
- Explain in language appropriate to patient and relatives
- Stop – Give time for information to “sink in”
- Ensure there are no misconceptions
- Communication - Liaison between hospital and community.

# Parallel planning:

- A key principle in providing good palliative care is parallel planning. This is the process of planning for ongoing care alongside planning for end of life care. It takes account of the often-unpredictable course of life-limiting conditions and involves making multiple plans for care, and using the one that best fits the patient and family's circumstances at the time.

(NICE, 2106)

# Poor prognosis letter to G.P.



# Quiz Time:





# Question 1:

What is a cardiac baffle?

- A. Intracardiac pathway / wall created from endogenous, synthetic or autologous pericardium tissue.
- B. A small window or hole in the patch used to create the intra-atrial tunnel in a Fontan operation.
- C. A procedure for an atrial arrhythmia.

## Question 2:

What lesion was the Fontan operation *initially* created for?

- A. Tricuspid Atresia
- B. Pulmonary Atresia
- C. Hyperplastic left heart

# Question 3:

What is the actual “Mortality age” of a 20 year old Fontan patient?

A. 64

B. 65

C. 66

# Question 4:

Name one of Choussat's ten commandments for a good Fontan operation?

- A. Normal Sinus Rhythm
- B. Competent Aortic Valve (AV)
- C. Competent Pulmonary Valve (PV)

# Question 5:

What anatomy requires a Fontan operation for survival?

- A. Single ventricle
- B. Double outlet
- C. All of the above

**Question 6:**

# Question 7:

When should we initiate discussion regarding “parallel planning” of care with a patient who has had a Fontan circuit?

- A. When the patient gets increase in severity and frequency of symptoms
- B. Wait for the patient to ask “Am I going to get better”? and then initiate conversation
- C. It is never too soon to start forward thinking and hope for the best but plan for the worst.

# References.

- Barbara J Deal. Marshall L Jacobs (2012).
- Hardiman, T (2013). British Journal of Cardiac Nursing. Vol 8 (7)
- ESC Guidelines – GUCH [www.escardio.org/guidelines](http://www.escardio.org/guidelines). Management of grown-up congenital heart disease (2012)





**KEEP CALM**

**and**

Thank you for your attention

**Any questions?**

**No? Great, bye!**



**Diolch**

**Yn**

Fawr

**Am**

**Wranddo**

