

SUPPLEMENTARY MATERIAL

Categories & questions taken forward to the workshop

Alternatives to a Fontan circulation

Can we define which patients would be best served by a Fontan circulation and which have a circulation which is balanced enough so that they do not require Fontan completion?

What are the alternatives to the current multi-stage surgical palliation?

How can the need for Fontan completion be delayed, perhaps indefinitely, to avoid the long-term progressive consequences of raised IVC pressure?

Can a Glenn shunt plus antegrade pulmonary flow provide adequate palliation without proceeding to Fontan completion?

Which patients would be better off going down a non-Fontan route?

How do the saturations, exercise capacity and life expectancy of patients palliated with a Glenn shunt plus antegrade pulmonary flow compare to that of Fontan patients?

Anticoagulation

What is the optimal anticoagulation regime for patients with a Fontan circulation eg. warfarin v aspirin v NOACs v none?

Does the optimal anticoagulation regime differ between patients, eg. by the type of Fontan - AP connection v Lateral tunnel v TCPC?

What are the risk factors for thrombosis following surgery for a single ventricle heart condition?

What is the optimal anticoagulation regime for patients with a Fontan circulation and atrial flutter?

What is the emotional impact of warfarin therapy on girls with a Fontan circulation going through puberty?

Arrhythmias

How can we reduce arrhythmia burden and does ablation/therapy improve symptoms?

Is ablation or amiodarone the best first line treatment for patients with a single ventricle who present with atrial flutter?

Does atrioventricular synchrony improve cardiac output and physical exercise in patients with Fontan circulation and sinus node dysfunction?

How common is sudden cardiac arrest in patients with a Fontan circulation?

How common are palpitations after Fontan completion?

Associated co-morbidities

What are the common co-morbidities in children with a single ventricle heart condition?

What are the outcomes in children with multiple co-morbidities?

Is there a correlation between single ventricle heart conditions and other 'unseen' illnesses, such as autism, ADHD, hypersensitivity, peripheral neuropathy, hypermobility, gastritis, coeliac disease?

What is the impact of syndromes or other co-morbidities on the quality of life of those with a single ventricle heart condition?

What are the non-cardiac morbidities in children with single ventricle heart conditions and what is the impact on parents and siblings?

Brain & neurodevelopment

What is the impact of a single ventricle heart condition on neurodevelopment?

What factors influence neurodevelopmental outcomes in children with single ventricle heart conditions?

Are there differences in brain development in the fetus with a single ventricle heart condition and if so, what is the impact on outcomes?

What is the impact of staged surgical repair on neurodevelopment?

Are problems with coordination more common in patients with a Fontan circulation and if so, why?

Exercise

What are the effects of regular physical exercise on well-being and long-term outcome in the Fontan circulation?

What are the benefits of exercise rehabilitation in well and deteriorating patients with a Fontan circulation?

What type of exercise is most beneficial in those with a Fontan circulation?

What is the effect of exercise on heart function in the Fontan circulation?

What is the ideal heart rate during exercise for those with a Fontan circulation compared with those with a two ventricle circulation?

What is the role of exercise plans during childhood in preventing heart failure later in life with the Fontan circulation?

Fontan failure

How can early failure and complications of the Fontan circulation be prevented?

What are the best markers of deterioration in patients with a Fontan circulation?

What is the main mechanism behind failure of the Fontan circulation?

How can we prevent late multi-organ dysfunction in the Fontan circulation?

How can late complications of the Fontan circulation such as protein-losing enteropathy (PLE) be prevented?

Heart function

How can ventricular function be best preserved in patients with a single ventricle heart condition?

What treatments can be developed for ventricular failure in patients with a Fontan circulation?

Do drugs such as ACE inhibitors, angiotensin receptor blockers, beta-blockers & aldosterone antagonists cause myocardial remodelling in the Fontan circulation?

What is the effect of exercise on heart function in the Fontan circulation?

What is the cause of diastolic dysfunction in the single ventricle?

What is the optimal strategy to preserve diastolic ventricular function in the Fontan circulation?

Living with a single ventricle heart condition

How can we best support children & young people with a single ventricle and their families in their day-to-day lives?

What are the frequencies of symptoms, limitations, physical and emotional quality of life with a Fontan circulation?

Which models of care, virtual ward and home monitoring programmes, best support infants, children and young people with a single ventricle heart condition at home?

How is emotional and behavioural development affected by having a single ventricle heart condition?

How can the complications of a single ventricle heart condition be better managed in the community?

What are the long-term social, psychosocial and other non-clinical outcomes of children with single ventricle heart conditions?

What lifestyle choices (eg. diet, exercise, occupation, physical location) best support the long-term health of patients with a single ventricle heart condition?

Management of the well-functioning Fontan circulation

How can the longevity of the Fontan circulation be prolonged?

How can organ function be optimised in patients with a Fontan circulation?

How does the Fontan circulation impact on other organ systems?

What factors best determine a well-functioning Fontan circulation?

What is the optimal interval for follow-up of patients with a Fontan circulation?

How often should follow-up imaging be performed in patients with a Fontan circulation?

What is the impact of pregnancy on patients with a Fontan circulation and how can outcomes be improved?

Outcomes of current treatments

How can we improve life expectancy and quality of life for patients with a Fontan circulation?

What are the current risk factors for death, reintervention and complications in patients with a single ventricle heart condition in the UK?

What are the long-term exercise and functional capacity of patients with a Fontan circulation and what influences outcome?

Is there a difference in outcome between systemic left and right ventricles in a single ventricle circulation?

Is bacterial endocarditis more common in patients with a single ventricle heart condition now that antibiotics are not routinely recommended prior to dental treatment?

What is the extent of morbidities in patients with a Fontan circulation eg. arrhythmias, functional, neurocognitive, developmental and psychiatric burden?

What outcomes matter to patients with a single ventricle heart condition and their families?

Quality of life

How can quality of life of those with a Fontan circulation be optimised?

What are the patient reported quality of life outcomes in single ventricle heart conditions?

What metrics are best to evaluate physical/neurodevelopmental and quality of life outcomes in those with a single ventricle heart condition?

What is the quality of life of adults with a Fontan circulation?

How does quality of life change over time in patients with a Fontan circulation?

What is the quality of life of the family of those with a single ventricle heart condition?

Surgery & perioperative care

How can perinatal risk stratification be used to identify those in whom the Norwood operation is futile?

How could technology be used to perform a biventricular repair in patients with a single ventricle heart condition?

Which modifiable perioperative factors can reduce mortality for the Norwood operation?

What is the optimal timing of the stages of surgical intervention?

Does post-operative NIRS monitoring reduce the incidence of adverse events in PICU?

Transplantation, mechanical support & stem cell therapy

Could stem cell therapy be used to grow a normal heart from the patient's own cells?

Could a patient's own cardiac stem cells be extracted in later years without open heart surgery and reinjected to improve cardiac function?

How can mechanical assist devices be developed to support the Fontan circulation?

Is there a role for stem cell therapy in the failing Fontan?

What are the alternatives to transplantation for the failing Fontan?