

Proposal of a UK & Ireland Single Ventricle Registry

Dear colleagues

Many of you kindly participated in the recent research priorities in single-ventricle heart conditions process (1). During and following that process, discussions with patients and their families and clinical teams caring for these patients have highlighted a wish to develop a UK and Ireland Single Ventricle Registry. We have seen the success of the Australia and New Zealand Fontan Registry in publications in a number of areas and the consensus following the research prioritisation process was that we should work together on developing a registry to address a number of the identified research priorities. With an average of 200 completed Fontan procedures and 100 Norwood procedures performed annually in the UK and Ireland, such a registry could rapidly accumulate data.

We propose that a registry be developed to include all congenital cardiac centres in the UK and Ireland, to collect data on single ventricle patients. We have agreed with Actelion Pharmaceuticals a one off non-restrictive grant to develop the registry based on costings delivered by the IT team at the Queen Elizabeth Hospital in Birmingham. The team have previously developed and continue host national and international registries and are familiar with the requirements of such multi-centre registries.

We have agreed additional funding for a launch meeting to discuss how this is developed and would wish to have all centres represented, ideally 2 representatives per centre and we are planning the meeting for Thursday 6th June 2019 in Birmingham; this is the day before our annual workshop meeting, which this year will also be on the single ventricle circulation. The charity Little Hearts Matter and the BCCA are supportive of this venture.

The registry would ideally allow data collection from fetal diagnosis to end of life and should address areas of interest highlighted in the prioritisation process (appendix 1).

The launch meeting will discuss the composition of a steering group and a publications sub-committee, the scope of data collected, data sharing agreements and long term aims.

The meeting will be coordinated by CFS events and will include overnight accommodation for participants out of area, and travel costs and will be held in central Birmingham. If you would like to attend this meeting please register here: <https://cfsevents.eventsair.com/uk-ireland-single-ventricle-registry/registration/Site/Register> or contact olivia@cfsevents.co.uk for further information.

Many thanks

Paul Clift

pclift@nhs.net

Reference:

1. Drury NE, Stoll VM, Bond CJ, Patel AJ, Hutchinson S, Clift PF, on behalf of the collaborators. Research priorities in single-ventricle heart conditions: a United Kingdom national study. *Cardiology in the Young* 2018 Dec 21:1-7.

Appendix 1. Identified research questions suitable for a UK and Ireland National Registry

1) **Associated co-morbidities**

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

2) **Brain and Neurodevelopment**

- a) What factors influence neurodevelopment outcomes in children with single-ventricle heart conditions?
- b) What is the impact of a single-ventricle heart condition on neurodevelopment?

3) **Fontan Failure**

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

4) **Management of the well functioning Fontan circulation**

- a) What factors best determine a well functioning Fontan circulation?
- b) What is the optimal interval for follow-up of patients with a Fontan circulation?
- c) What is the impact of pregnancy on patients with a Fontan circulation and how can outcomes be improved?

5) **Surgery and preoperative care**

- a) Which modifiable perioperative factors can reduce mortality for the Norwood operation?
- b) How can perioperative risk stratification be used to identify those in whom the Norwood operation is futile?

6) **Transplantation, mechanical support and novel therapies**

- a) What are the roles of novel therapies in the failing Fontan?
- b) What are the alternatives to transplantation for the failing Fontan?