

The Australian & New Zealand Fontan Registry Newsletter

Welcome to the 2018 Fontan Registry Newsletter!

Thanks to your enormous support, we continue to break ground in research to get the best possible life to those with a Fontan circulation.

This year is very special because it marks 50 years since the first Fontan procedure was done (first operation in 1968 and first made public by publication in 1971) and the passing of its creator, Professor Francis Fontan.

The Australian and New Zealand Fontan Registry continues to drive Fontan research in our countries and internationally. We have this year made many new discoveries. Our research is starting to clarify what happens in the body when this operation has been done. We still do not understand everything, but have a clearer picture on the examinations that are necessary to make sure that all have the best possible health.

We would like to thank all of those who have participated in our cross-sectional Functional Outcomes study. We have completed this year our data collection and have accumulated a vast amount of information. We are now crunching this data and it is promising already to give very exciting and novel results!

We have also more and more realised that exercise is important. We are trying to investigate how to best achieve a good exercise capacity with a Fontan circulation. (If you are willing to start a new exercise regimen, do not forget to see first your cardiologist!).

This year also, in our region and throughout the world, several trials have been started to investigate what best medications to take when one has a Fontan circulation in a way that has never been seen before.

I have the privilege to follow very closely all the research done in this space and I promise you that things are changing fast. I could see this year to which degree everybody in the world is accelerating this research. There is now a sense of urgency of getting results that was not there a few years ago.

Thank you to all of you ... and watch this space, a lot of exciting developments will come forward in the next few years!



*Prof Yves d'Udekem
Australian and New Zealand Fontan Registry Founder and Chair*



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UPDATE ON FONTAN REGISTRY

Participation in the Registry has grown steadily to now include a total of 1,574 participants. This is matched by a steady increase in the number of Fontan procedures per year since the 1970's.

The graph below shows the rapid growth in the number of patients with a Fontan circulation. At the end of 2017, 137 patients had an Atriopulmonary connection (AP); 253 had a Lateral Tunnel (LT) and 1053 patients had a Extracardiac Conduit (ECC).

In regards to the number of Fontan operations conducted, 46 new Fontans were recorded in the Fontan Registry at the end of 2017. The average age of participants is 18 years, with just over half of the participants (57%) being male. The average age at time of Fontan operation is currently 5.7 years (ranges from less than 1 year old to 41 years old at the time of operation).

The ongoing growth and size of the Fontan population is important to track as it helps us focus our research, and advocate for the best long-term health and wellbeing outcomes with policy makers.



Download our annual report here:
<https://www.fontanregistry.com/annual-reports>

Australian & New Zealand
FONTAN REGISTRY:
REPORT 2017

August 2018

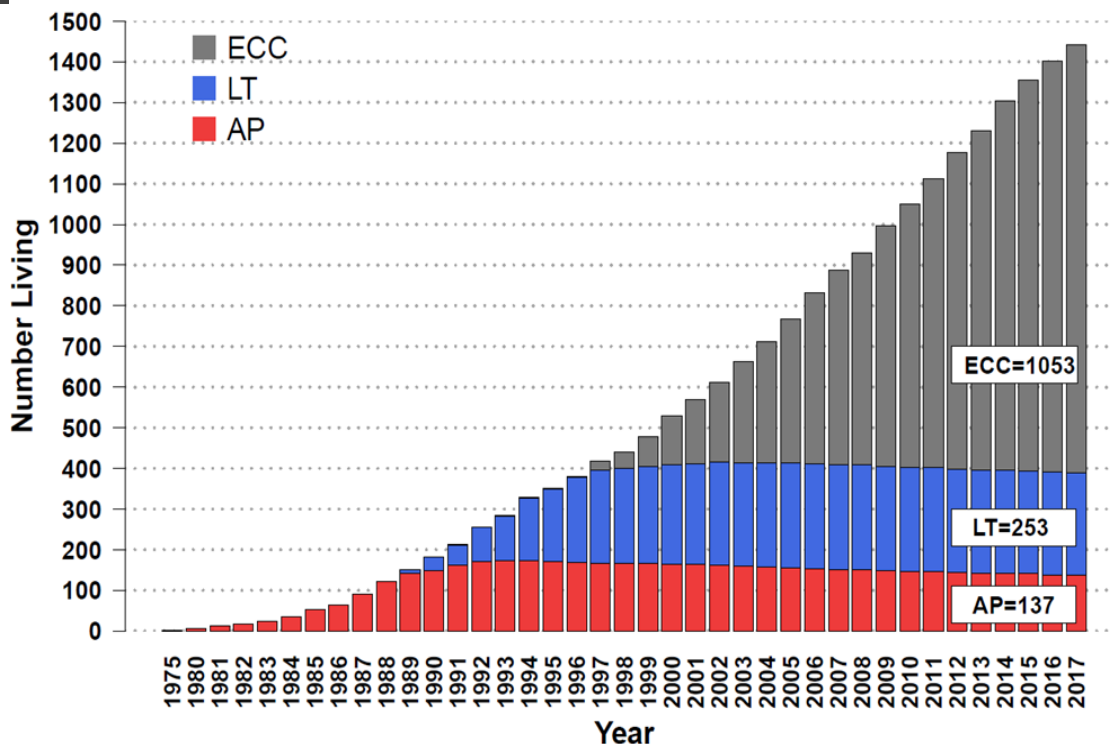


Figure 1. Number of Registry participants alive with a Fontan circulation (N = 1,443)

ANZFAC UPDATE



AUSTRALIAN &
NEW ZEALAND
FONTAN ADVISORY
COMMITTEE

As Christmas nears and 2018 draws to a close we want to thank you, the Fontan Community, HeartKids Ltd, Heart Kids NZ, and the ANZ Fontan Registry, without whom what we do and are trying to achieve, would not be possible.

2018 has been a busy year for ANZFAC as we became a subcommittee of HeartKids Limited, drafted our advocacy document, recommenced Fontan Carers Groups, launched Creative HeART, continued with our Fontan Personal Stories and Q&A projects on Facebook, fundraised for the Fontan Registry with *Footy for Fontan*, continued to plan our increased presence in and representation from New Zealand, provided feedback on the National CHD Action Plan, presented at Fontan Education Day in Adelaide, explored involvement in the HeartKids Peer-to-Peer Mentor Program, provided representation on the Fontan Registry Steering Committee, and fulfilled an advisory role on select projects with the Fontan Registry and others.

This year we were also delighted to welcome two new committee members. Holly Saunders and Jaden Dixon, who have brought a highly valued youth perspective. We also welcomed our new Deputy Chairperson Patrick McConville, who has brought to the role enthusiasm, personal insight of an adult with a Fontan, leadership, knowledge and a true sense of community. We thank our former Deputy Chairperson Ingrid King for all her incredible work, dedication and passion to make a difference in the lives of people with a Fontan circulation and their families. We are very happy that she has remained on the Committee.

If you would like to find out more about who we are, what we do, our projects, or you would like to be involved in some way, you can reach us via the details below.

Email: anzfac@heartkids.org.au

Facebook: www.facebook.com/ANZFAC/

Website: <http://www.heartkids.org.au/ANZFAC>

ANZFAC would like to wish the Fontan community a Merry Christmas and a safe, healthy and happy holidays. We look forward to a fabulous and productive 2019.

Rachel Maree
Chairperson, ANZFAC



Pat McConville (*Deputy Chairperson*)
presenting at Education Day 2018



Fontan Carers Group Perth (November
10, 2018)





OUR PROJECTS

Rubato Clinical Trail (*Currently Recruiting*)

The Australian and New Zealand Fontan Registry is running a randomised controlled trial. It is a prospective, multi-centre, double-blind, randomised, placebo controlled trial. The trial is sponsored by Actelion (under Johnson & Johnson). The study is to assess the efficacy and safety of Macitentan in Fontan-palliated adult and adolescent patients.

The primary outcome is to determine if there is any change in peak oxygen uptake, and if it can help improve the ability of people with a Fontan circulation to exercise.

Participation in the study lasts for about 1 year and involves several visits to various study sites around Australia and New Zealand.



ACE Inhibitor Cessation Trial (*Currently Recruiting*)

ACE inhibitors are a group of medications that relax blood vessels and lowers your blood pressure. The most commonly prescribed which you might be familiar with are Perindopril, Lisinopril, Enalapril, Captopril.

The ACE Cessation study is looking at the impact of angiotensin inhibitors in children and adults with a Fontan circulation and normal heart contraction. The evidence is showing that ACE inhibitors are of benefit in a failing 2-ventricle heart, but there is no evidence to suggest they are of benefit in the Fontan circulation.

The project aims to demonstrate that stopping ACE inhibitors will not result in a significant decline in peak exercise capacity, or cardiac function over the 12-month study period.

We hope that through this research, we can demonstrate that these medications are not necessary in particular circumstances. It may also help to protect people with a Fontan heart from adverse drug reactions and the burden of life-long medication use.

FAN Trial (Fontan Associated Nephropathy)

The FAN trial is a randomised–controlled trial which is assessing the effect of a drug called Perindopril on Fontan-Associated Nephropathy.

Nephropathy is a medical term that refers to damage to the kidneys, resulting in increased levels of protein in the urine and/or loss of kidney function over time.

Participants involved in this study will take the study drug or placebo for 6 months. There will also be 4 visits to the hospital and a phone call. During these visits we will carry out a few tests to assess your progress and keep track of your health.

We hope that through this research, we can determine if Perindopril is an effective treatment for nephropathy for those with a Fontan circulation.



Analysis of factors associated with exercise function in the Fontan physiology

This study is to determine what factors are the most important predictors/associations for a superior Fontan circulation (where your peak oxygen and peak work is greater than 80% predicted). The main focus will be on exercise and to determine if increased early life physical activity levels play a role in superior function late after Fontan palliation.

We will explore the family dynamics and social influences surrounding physical activity between the study and control group.

Participants will complete a series of exercise questionnaires over the phone at a convenient time (30 min)

We hope through the analysis of the results obtained, we can determine what factors, such as impact of family, culture and social influences that play a role in an individuals involvement in sport and exercise.



If you would like to participate in any of the above projects, please don't hesitate to contact us:

E: info@fontanregistry.com

Or

T: +61 3 9345 6161





WHAT IS RESEARCH?



With such a great number of trials being carried out at the Fontan Registry, it is important to know what research is. How the Fontan community will benefit from the research, and how YOU can get involved and contribute to research.

- Research is defined as the creation of knowledge and/or the use of existing knowledge in a new and creative way so as to generate new concepts, methodologies and understanding.
- Through our research we aim to improve the lives of people living with a Fontan circulation.
- All new research projects start off with a clear research question that we want to answer.
- The main way to answer a research question is to look at the data collected.
- Data can be collected through various methods including, looking at data already in the Registry, or gathering new data.
- The gathering of new data can happen through surveys, through medical tests.
- One main method we use for gathering this data is by conducting clinical trials.
- The results from there trials will be shared with the wider Fontan community and in peer-reviewed journals.

Why should YOU participate in research?

- It important that you participate in research as the field of Fontan research is still relatively new and we are trying to expand our knowledge to help those living with a Fontan circulation and those yet to come.
- In research we need large numbers to participate in projects so that we can draw accurate conclusions.
- By participating in research, you might also learn some new information about yourself, including up to date clinical information.
- It can also just FEEL GOOD to participate. By doing your little bit, you are helping move science forward and contributing to the knowledge base on Fontan outcomes.

YOUR participation can help you, but also importantly so many other people.



FONTAN RESOURCES

We recently added a new section to our Fontan Registry website (<https://www.fontanregistry.com/>) called **Fontan Specific Resources**.

In collaboration with doctors, parents and Fontan patients we were able to develop a document that outlines the most commonly used medical tests. We hope that this can be used as a valuable resource for Fontan patients and their parents to get a better understanding of the tests that patients will encounter throughout a typical year. A snippet of what the document contains can be found below.

We also have a document with an easy to follow glossary, which lists commonly used terms that you may have heard being mentioned at an appointment or clinical review.

Link to both documents:

<https://www.fontanregistry.com/fontan-specific-resources>

Oxygen Saturation (Pulse Oximeter)

Most people have an oxygen saturation $\geq 96\%$. Many people with a Fontan circulation will have a slightly lower reading – it is important for you or your parents/carers to know what the usual reading for you is.



The oxygen saturation (saturation of red blood cells with oxygen) is measured most often using a probe (pulse oximeter). The probe can either sit on a finger, a clip that goes on the ear, or a membrane that is taped onto the forehead (depending on the age or size of the person). The pulse oximeter makes a pinging sound (alarm) that lets doctors or nurses know of changes in oxygen levels. It is very useful particularly during surgery, and staff are trained to leave the alarm on, even if you are awake and well.

Sometimes, your doctor may want to know what happens to your oxygen saturation when you exercise. You may be asked to do an exercise test (usually using a treadmill or bicycle, or sometimes just walking) with a pulse oximeter on.

Blood Pressure

Monitoring the blood pressure is very important so that it can be maintained within a healthy range. Abnormal blood pressure readings, either high or low, may need to be addressed by your doctor. Blood pressure is measured using a special cuff that is inflated on the arm for a few seconds. The pressure of this cuff around the arm needs to be quite high to accurately measure the pressure in the blood vessels. Some people may find the pressure of the cuff uncomfortable. If you/your child finds blood pressure measurement distressing you should let your doctor know.





FONTAN RESOURCES

People with a Fontan circulation will have blood tests done. For adults this is usually yearly and in children this may be performed less frequently.

Some of the blood tests that you/your child might have are:

Full blood count and iron study- many people with a Fontan circulation may have a slightly low oxygen level. A full blood count and iron studies is done to make sure the person has enough iron to match their red blood cell making needs (the red blood cells carry oxygen in the body, and iron is required to make red blood cells).

Liver function tests, urea and electrolytes – these tests check how the liver and kidneys are working. Liver problems can arise in the Fontan circulation, and although why and how to monitor this is not well understood, blood tests form part of the regular checks. Although reduced kidney function has been noted in the Fontan population, it is unclear whether this is progressive. Testing the kidney function is a good screening and monitoring test.



Thyroid function and uric acid - in the general population thyroid issues are relatively common and can be a cause for tiredness or palpitations, so they are often included in blood tests. Sometimes people who have low oxygen levels have a higher rate of cells in their bodies being made and destroyed. This causes uric acid levels in the body to rise - uric acid crystals sometimes form in the joints and cause a painful condition called gout. Gout can be well treated with medication (usually a pre-venter medication).



Blood tests are performed with a needle. If you/your child find having blood taken uncomfortable, have a chat to your doctor or laboratory about the possibility of having some numbing cream applied to the skin 1 hour before.

If you/your child are on warfarin (an anticoagulant) then the blood thinning level (INR) needs to be checked regularly (this can vary from a few times a week if it is unstable, to 4-6 weekly if the level is stable) with an INR test. People on warfarin will have an INR range to aim for – for most people this is between 2.0 and 3.0. It is essential you check with your doctor what the INR range for you/your child needs to be, and that an INR test is done as frequently as advised. It is also important to know if you/your child are taking warfarin that the INR levels can be affected by foods, medications, and almost all antibiotics. When starting a new medication, make sure that the doctor and pharmacist know that you/your child are on warfarin and follow their advice on INR checking (usually INR is checked on day 3 of the new tablet). Sometimes, people on warfarin self-manage their INR checking at home with a finger prick test machine.

Echocardiogram (ECHO)

An echocardiogram, or “Echo”, can be used to provide a picture of the heart chambers and valves and check how well they are working. An Echo uses sound waves to form a moving picture of the heart and major blood vessels, the sound waves are converted to a picture by a computer on the Echo machine. The sound waves are recorded by a hand-held wand, known as a transducer that is placed on the chest.

The Echo may be performed by a technician who works with your doctor, known as a cardiac sonographer. Before the test you/your child may be asked to remove clothing from the waist up and wear a gown. To perform an Echo, a small amount of gel is placed on the end of the transducer to allow better contact with your/your child’s skin and provide a clearer picture. Three ECG electrodes are often put on the chest in order to record the heart’s electrical activity during the test. The technician will move the transducer to different parts of the chest to get a complete picture of the heart and blood vessels. You/your child may be asked to change positions several times during the test and briefly hold your breath to provide a clearer picture of your heart. The technician may take measurements of the size of different heart chambers, valves or blood vessels.



You/your child should not feel any pain or discomfort during an Echo, the gel may feel a little cold though and slight pressure may be felt from the transducer placed on the chest. The sound waves used in Echo are not known to lead to any long term harmful effects.

Electrocardiography (ECG)

An electrocardiogram, usually abbreviated to ECG, records the electrical signals from the heart and whether the rhythm of the heartbeats is steady or irregular. It also gives some very basic information about the heart structure.



To record the ECG, twelve little sticky patches called ‘electrodes’ are put on the chest, arms and legs. Your skin may need to be shaved to help the electrodes stick. The stickers are connected to an ECG recording machine that records your/your child's heart electricity into wavy lines which are printed onto paper. The test is safe and only takes a few minutes in total. The ECG records electricity and does not affect the heart in any way.



FONTAN EDUCATION DAY 2018- ADELAIDE

Our 5th annual Fontan Education Day was held on 9th September 2018 at SAHMRI. It was a beautiful location for our Education Day and the event was attended by 150 Fontan participants, their families, cardiac specialists and researchers. As with previous Education days, this was a great opportunity for patients and their families to hear from experts in the field and importantly to share patient insight and lived experiences. Families were able to connect with each other and it allowed them to hear about the latest research and results from the Fontan Registry.

Highlights of the day included: a thorough outline of the *Fontan procedure*, a look at *the advances in technology and the International Fontan Interest Group*, *Pregnancy and Fontan*, *Exercise and Fontan*, *A parent and patient story*, *Dentistry and Fontan* and *Neurodevelopment*. There were in-depth break-out sessions which gave patients and their families a chance to explore topics further and engage with cardiologist and researchers.

The day was well received, with lots of positive interactions and sharing of experiences. Thank you to our wonderful Adelaide team and the planning committee for their exceptional efforts in hosting another great event! We look forward to seeing you in Melbourne next year.

Here is a link to the videos of the day:

<https://www.fontanregistry.com/seminar-videos-2018>



Announcement:
Fontan Education Day MELBOURNE
Date: 14th Sep 2019



STAY CONNECTED

Please follow our updates **on our Facebook site:**
<https://www.facebook.com/ANZfontanregistry>
to connect with other Fontan families like you.



The **Fontan Youth Support Network** is a specific online support group for young people aged 15-25 (This is a closed group). For more information please visit:

<https://www.fontanregistry.com/fontan-youth-support-network>

To support the Fontan Registry and ongoing research, donations can be made directly to the Fontan Registry (we now also support regular giving and all donations are tax deductible). To be one of our **#fundraisingheroes** please visit our site:

<http://www.fontanregistry.com/donations>

Your continued support is much appreciated!!



“Giving is not just about making a donation.

It is about making a difference.”
#researchsaveslives

HAVE YOU MOVED HOUSE?

New mobile? New email address?

Please help the Fontan Registry to keep our records up to date by letting us know of any updates/changes to your contact details.



To update your details visit:
<http://www.fontanregistry.com/participate>



OUR ACTIVE FONTANS

A whole life with half a heart





SEASONS GREETINGS



From everyone at the Fontan Registry, we wish all the Fontan participants, their families and friends a safe and happy festive break.

We would also like to take the time to thank everyone involved in the Fontan Registry team for their hard work and dedication this year.

Best wishes to All for a wonderful 2019!



The Fontan Registry Team at Murdoch Children's Research Institute

