

# The Australian & New Zealand Fontan Registry Newsletter

## Welcome to the 2017 Fontan Registry Newsletter!

There has been a perceptible change in the sphere of Fontan research in the last year, and this change has been driven by research done within the Australian and New Zealand Fontan Registry. It is now becoming obvious that everyone is realising that the Fontan population throughout the world is becoming larger than anybody would have ever expected, and that our community has to support them to have the best possible life.

As more data are now being gathered, we now have an estimate of the world population of those with a Fontan circulation of around 60,000 people! From our Australian and New Zealand population-based data, 40% of them are adults above 18 years of age. Most of them are working. People are realising that we do not know enough to take care of them all properly and initiatives are underway.

A promising international collaboration has been created (the IFIG-International Fontan Interest Group). For the first time, now there are several clinical trials of medication in the world that specifically target the well-being of those with a Fontan circulation. Nothing is obvious yet, but a lot of efforts are being made to provide an artificial heart solution. All these efforts have become a reality as a result of the good work of the Fontan Registry.

**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*

## 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>



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[www.fontanregistry.com](http://www.fontanregistry.com)  
[info@fontanregistry.com](mailto:info@fontanregistry.com)



## UPDATE ON FONTAN REGISTRY NUMBERS

Participation in the Registry has grown steadily to now include a total of 1,524 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 2016, 133 patients had an Atriopulmonary connection (AP); 259 had a Lateral Tunnel (LT) and 995 patients had a Ex-tracardiac Conduit (ECC).

In regards to the number of Fontan operations conducted, 52 new Fontans were recorded in the Fontan Registry at the end of 2016. The average age of participants is 19 years, with just over half of the participants (56%) 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.

## NEW RESEARCH

### Pregnancy and Fontan

This year we explored the potential long term impact of pregnancy for women with a Fontan circulation.



We looked at the big picture outcomes in women, who have/have not had a pregnancy. We identified 30 women who had 46 pregnancies that went beyond 20 weeks gestation. Typically, the women who had a pregnancy were older. They also appeared to be healthier, with fewer medical events pre-pregnancy.

For these women, the data suggests an increase in thromboembolic events after pregnancy. We do not know whether this is related to pregnancy, or the fact that these women were older and more years post Fontan.

As this is a small group of women with a limited amount of follow up post pregnancy the study has highlighted the need for matched data, to allow us to answer questions about possible long term effects. This idea will be the focus of an exciting international collaboration planned to start in 2018.

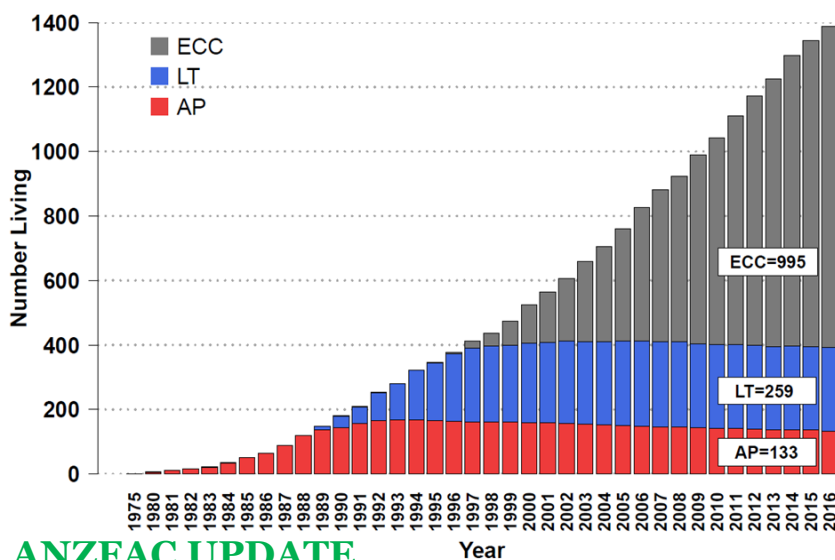
**Women who are considering pregnancy should discuss a plan with their cardiologist well in advance of getting pregnant.** It is important to be closely followed before, during and after pregnancy for best outcomes for mum and baby.

Here is our link to the published review:

<http://journals.sagepub.com/doi/pdf/10.1177/1753495X17737680>

If you have any details regarding your pregnancy and birth history that you think the Registry may not know, please contact us and allow us to update your information.

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



## ANZFAC UPDATE

ANZFAC would like to thank the ANZ Fontan Registry, HeartKids Australia and HeartKids New Zealand for their continued support throughout 2017. We'd also like to thank the Fontan community and we look forward to making some exciting announcements in the near future regarding ANZFAC, our projects and events for 2018.



You can stay up to date on our activities and contact us via, facebook: <https://www.facebook.com/ANZFAC/>

webpage: <https://www.fontanregistry.com/anzfac>

email: [anzfac@fontanregistry.com](mailto:anzfac@fontanregistry.com)

On behalf of ANZFAC we wish you all a Merry Christmas and a safe, healthy and happy holiday season!

Rachel Maree & Ingrid King

Chairperson & Deputy Chairperson

## LIVER RENAL PROJECT

Thanks to 150 of you, we have now completed our cross-sectional study of the impact of the Fontan circulation on liver and kidney function. This study is one of the most detailed to have seen the light of day so far. As we expected, we found that high pressures in the veins put pressure on these two organs. During the second decade after Fontan, it seems that half of those with a Fontan circulation would have some abnormalities of the liver and the kidney. But, these abnormalities at this stage are very mild and we have not seen much serious adverse consequences in our population of Australian and New Zealand Fontan patients. It is at least much less than what was expected from research done in other parts of the world.

Our conclusion is that in the second decade after Fontan, we should start monitoring these two organs, but if they will have an impact, it will be more likely to be later in the third and fourth decade after Fontan.

## OTHER RESEARCH PROJECTS UPDATE

People with a Fontan are known to have an increased chance of getting a blood clot. For this reason, patients are usually prescribed aspirin or warfarin to slow down the time it takes for blood to clot.

As these medications work in different ways, they have different side effects. One side effect is the potential reduction in bone mineral density (how dense and hence how strong your bones are) in patients receiving long-term warfarin treatment.

Early data from the Aspirin versus Warfarin study in the Fontan population has shown that participants receiving warfarin typically had lower bone mineral density than their aspirin counterparts. In July, these findings were presented at the *International Society on Thrombosis and Haemostasis Congress* in Berlin.

Although more analysis is required before recommendations on aspirin or warfarin treatment can be made, there are a number of general recommendations for children who are receiving long-term warfarin therapy. To improve their bone mineral density, it is recommended for children to:

- Eat 2-3 serves of dairy a day OR supplementation with calcium
- Take vitamin D supplementation at least throughout winter
- EXERCISE (as appropriate) - weight bearing best!

Weight bearing exercise (exercise done while on your feet so you bear your own weight). For example: **brisk walking, jogging, skipping, basketball / netball, tennis, dancing, stair walking**

- Ensure puberty is reached at an appropriate time (please discuss this with your doctor)

**Remember** always talk to your **cardiologist** about what's right for you!!



### Aspirin vs Warfarin??



## TRANSITION:

### MOVING TO ADULT CARE

Transitioning to adult healthcare is a crucial period for congenital heart disease (CHD) patients. It is essential they are followed closely as they are at a higher risk of adverse events.

We explored the readiness for transition to adult care of adolescents with a Fontan circulation and their parents. 17 adolescents aged 15-18 years and 15 of their parents were given questionnaires to complete at their first transition clinic.

The results showed that adolescents had poor knowledge about their Fontan circulation and 41% had a poor understanding of the purpose of their medication/treatment. They felt less comfortable talking about sensitive adolescent issues, in particular, emotional wellbeing. Parents also reported high levels of anxiety around their children transitioning to adult health care services.

These findings highlight the need for programs that focus on all aspects involved with transitioning, including parent involvement, a health system that is youth-friendly and a focus on emotional wellbeing.



## FONTAN EDUCATION DAY 2017 - BRISBANE

Our 4th annual Fontan Education Day was held on 9th September 2017 at Lady Cilento Children's Hospital, Brisbane. This 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 'who, what, where and why?'*, *Fontan through the ages*, *Pregnancy and Fontan*, *A family perspective*, *Moving to adult care*, and *Active living for the Fontan heart*.

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 Brisbane team and the planning committee for their exceptional efforts in hosting another great event! We look forward to seeing you in Adelaide next year.

**Here is a link to the videos of the day:**

<https://www.fontanregistry.com/seminar-videos-2017>

**Announcement:**  
**Fontan Education Day Adelaide, Date: 8<sup>th</sup> September 2018**

### SEASONS GREETINGS

We wish all the Fontan participants and families a safe and happy festive break!

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

Best wishes to All for a wonderful 2018!



### STAY CONNECTED

Please follow our updates **on our Facebook site:**

<https://www.facebook.com/ANZfontanregistry>

to connect with other Fontan families like you.



Our recent addition to the Fontan Registry website is the **Fontan Youth Support Network**. If you are aged between 15-25 and have a Fontan circulation, this support network is just for you. For more information 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). To be one of our **#fundraisingheroes** please visit our site:

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



Your continued support is much appreciated!!