



# Australian & New Zealand FONTAN REGISTRY: *ANNUAL REPORT 2019*

September 2020



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## EXECUTIVE SUMMARY

The Australian and New Zealand Fontan Registry was created in 2009 as a population-based repository of Fontan patients' data. It aims to facilitate a greater understanding of these patients' short- and long term health outcomes, as well as factors which can be modified to improve their health and wellbeing.

This report focuses on data gathered up to the end of 2019, and was extracted from the database on the 10<sup>th</sup> of August 2020.

Participation in the Registry has grown steadily to now include a total of 1,682 participants. This is matched by a steady increase in the number of Fontan procedures per year since the 1970's. The average age of participants in the Registry is 19 years and just over half the participants are male (58%), and the average age at time of the Fontan procedure was 5.6 years.

The most common primary diagnosis for participants in the Registry included tricuspid atresia (22%), double inlet left ventricle (16%), double outlet right ventricle (14%), and hypoplastic left heart syndrome (HLHS) (13%). The main type of Fontan procedure performed on patients was Extracardiac Conduit (ECC) (69%) followed by Lateral Tunnel (LT) (17%), and the Atriopulmonary Connection (AP) procedure (14%). However, this picture has changed significantly over time with Atriopulmonary connection (AP) performed more commonly prior to 1990, lateral tunnel (LT) performed more often in the 1990's and the majority of the more recent Fontan procedures being Extracardiac Conduit (ECC).

With regards to medication, anticoagulants are most frequently noted in cardiologist review letters (data source). The majority of the patients in the Registry on which follow-up data is available in the preceding 2 years ( $N = 1028$ ), are on Aspirin (48%) or Warfarin (45%).

The report concludes by looking at the survival, arrhythmias and risk factors highlighting that there is no sudden decrease in survival in the early decades after Fontan surgery.

## FINDINGS

### RECRUITMENT

The findings presented in the current report concern Registry participants who had their Fontan operation up to the end of 2019. Data was extracted from the Fontan Registry on 10th August, 2019. As of this date, 1,682 eligible participants had been entered into the Registry (Table 1).

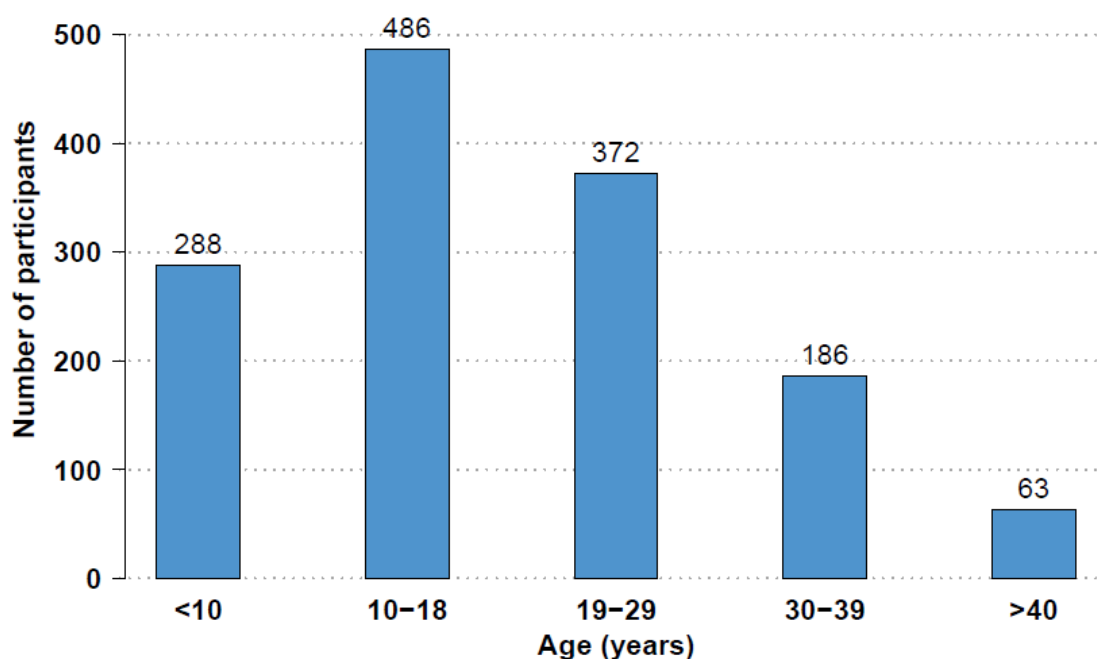
**Table 1 Number of Fontan Registry participants by current State/Country and enrolment status**

<i>Enrolment Status</i>	Total
Consented <i>(not exited)</i>	1395 (100)
Waiver of Consent <i>(uncontactable; hospital records only available)</i>	53 (100)
Transplanted*	35 (100)
Taken Down*	11 (100)
Deceased*	115 (100)
Not Consented / Other <i>(e.g., moved overseas)</i> <sup>^</sup>	52 (100)
Refused	17 (100)
Withdrawn	4 (100)
<i>Total Participants</i>	<i>1682</i>

Note: Figures in parentheses are row percentages. \*Denotes first exit point from Registry; Total deceased = 125; <sup>^</sup>includes a total of 21 international patients, 13 patients who have moved overseas and 18 patients not consented to date

### COHORT DESCRIPTION

The mean current age of alive and consented participants (N=1395) is 19.4 years ( $SD=10.5$ , range=1.8-55.5 years), with 52% under 18 years, and slightly more than half (57%) are male. The mean age of participants at the time of their Fontan operation was 5.6 years ( $SD=3.7$ , range= 0.3-40.9 years). The mean age at Fontan conversion operation (typically from an Atriopulmonary Connection to an Extracardiac Conduit) was 25.2 years ( $SD= 8.8$ , range= 7.4 - 45.6 years). The current age of the participants is shown in Figure 1.



**Figure 1 Current age of alive and consented Fontan Registry participants (N = 1395)**

The participants' primary diagnosis is shown in Table 2. Overall, the most common diagnoses are for *Tricuspid Atresia* (21.3%), *Double Inlet Left Ventricle* (16.1%), *Double Outlet Right Ventricle* (13.9%) and *Hypoplastic Left Heart Syndrome* (HLHS) (13.0%).

**Table 2 Primary diagnosis**

<i>Primary Diagnosis</i>	<i>n (%)</i>
Tricuspid Atresia	359 (21.3)
Double Inlet Left Ventricle (DILV)	271 (16.1)
Double Outlet Right Ventricle (DORV)	233 (13.9)
Hypoplastic Left Heart Syndrome (HLHS)	219 (13.0)
Pulmonary Atresia with Intact Ventricular Septum	142 (8.4)
Unbalanced Atrioventricular Septal Defect	130 (7.7)
Congenitally Corrected Transposition of Great Arteries (ccTGA)	109 (6.5)
Pulmonary Atresia with Ventricular Septal Defect	35 (2.1)
Ebstein's Anomaly	16 (1.0)
Other	154 (9.2)
Missing	14 (0.8)
<i>Total</i>	<i>1682 (100)</i>

Information on the type of Fontan is available for 1,666 operations. The most common type of Fontan operation was the Extracardiac Conduit (69.1%), followed by Lateral Tunnel (17.2%) and the Atriopulmonary Connection procedure (13.7%). More than one-third of Fontan operations were fenestrated (38%).

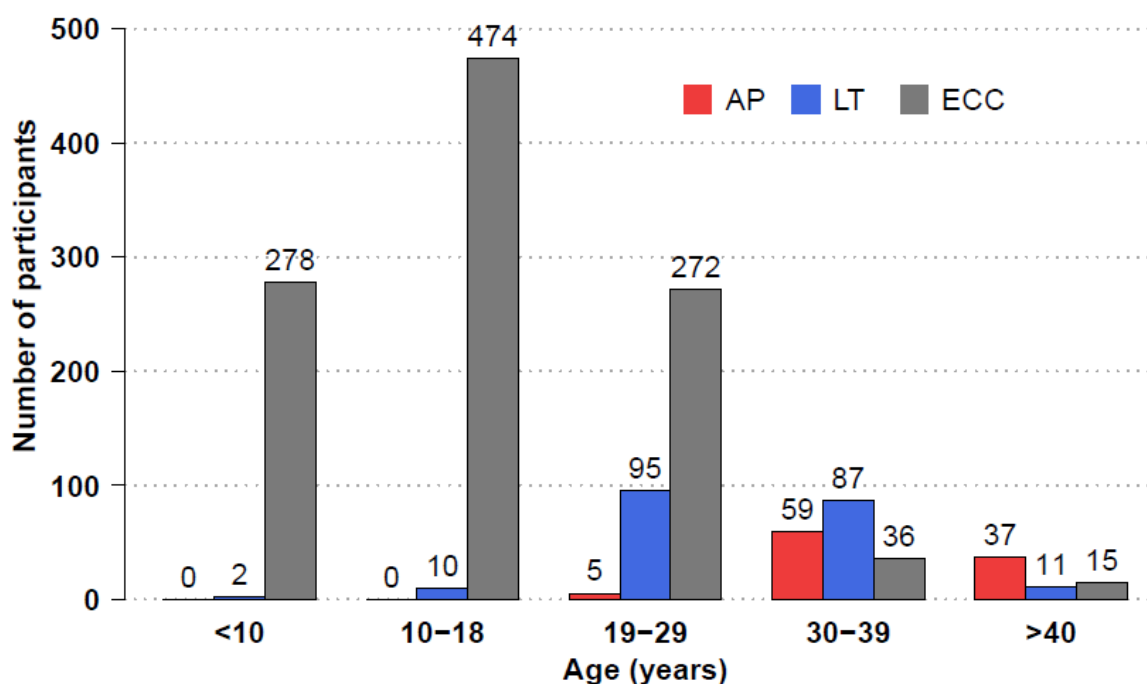
There has been a significant change in the type of Fontan operation over time with Atriopulmonary being most common prior to 1990, Lateral Tunnel common in the 1990s, and almost all recent Fontan operations being Extracardiac Conduit (Table 3).

**Table 3 Type of Fontan operation by date**

Year of Fontan	Fontan Procedure Type			Total
	Atriopulmonary	Lateral Tunnel	Extracardiac Conduit	
1975-1989	181 (95.3)	9 (4.7)	0 (0.0)	190 (100)
1990-1999	48 (13.5)	242 (68.0)	66 (18.5)	356 (100)
2000-2009	0 (0.0)	28 (5.4)	495 (94.6)	523 (100)
2010 -2019	0 (0.0)	7 (1.2)	590 (98.8)	597 (100)
<i>Total</i>	229 (13.7)	286 (17.2)	1151 (69.1)	1666 (100)

Note: Figures in parentheses are row percentages.

Figure 2 shows the age group of Fontan Registry participants broken down by the type of Fontan circulation the patient is living with (i.e., includes conversion to Extracardiac Conduit).



Note: AP=Atriopulmonary, LT=Lateral Tunnel, ECC=Extracardiac Conduit

**Figure 2 Current age of participants by type of Fontan operation (includes Conversion) (N = 1381\*)**  
 (\*14 missing Fontan type have been omitted)

Figure 3 shows the number of Fontan operations conducted per year as recorded in the Fontan Registry with 64 new Fontan operations in 2019. It shows the growth in this procedure over time with the last 10 years seeing an average of 61 operations per year.

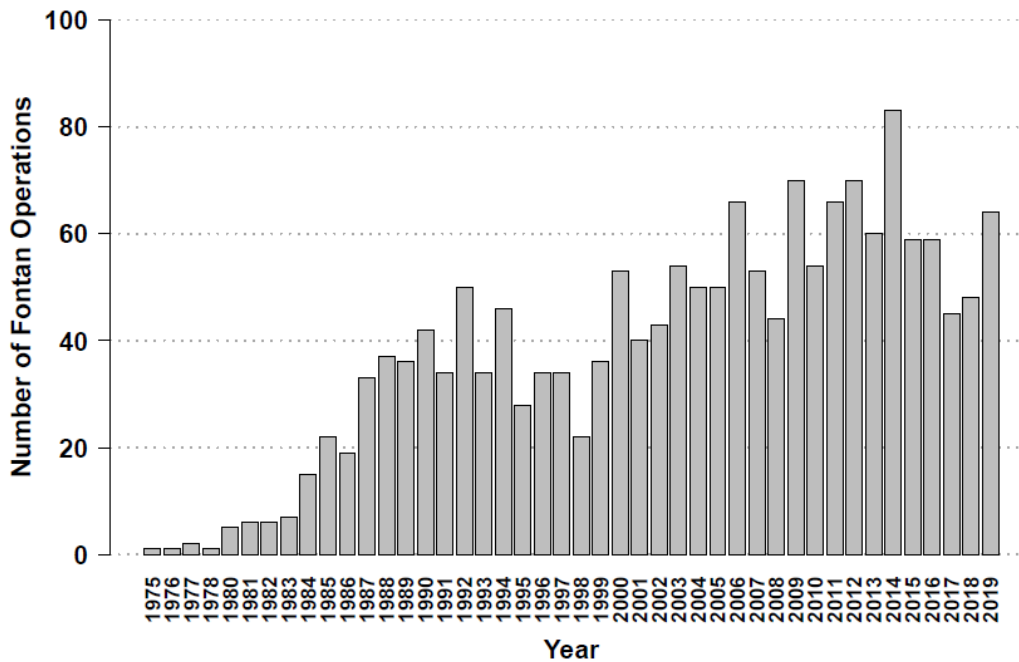


Figure 3 Number of Fontan operations per year

Figure 4 shows the rapid growth in the number of patients who are living with a Fontan Circulation. After taking into consideration those who had a conversion from an Atriopulmonary Connection (AP) Fontan, at the end of 2019, 120 patients were living with an Atriopulmonary Connection (AP); 247 were living with a Lateral Tunnel (LT); and 1137 patients were living with an Extracardiac Conduit (ECC).

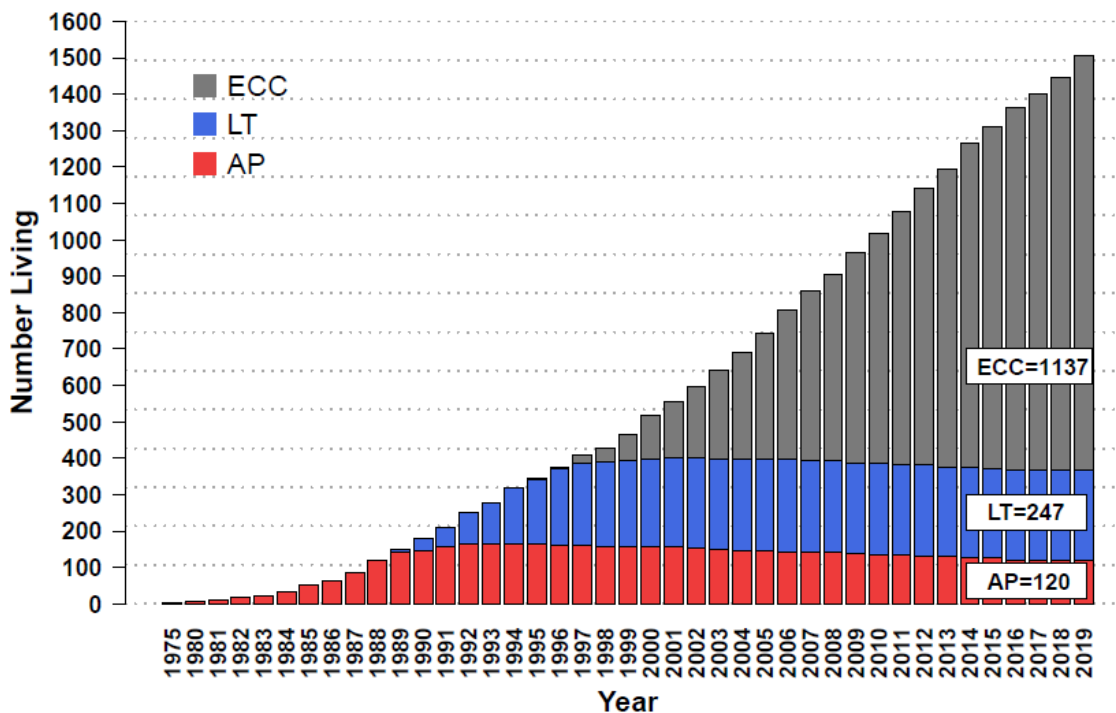


Figure 4 Number of Registry participants alive with a Fontan circulation (N = 1504)

## MEDICATIONS

Table 4 presents data on the use of anticoagulation medication among the 1028 consented participants as recorded on the most recent follow-up within the previous two years. Within this group of participants, Aspirin was the most commonly used anticoagulant, with 48% reporting the use of this medication followed by Warfarin with 45%.

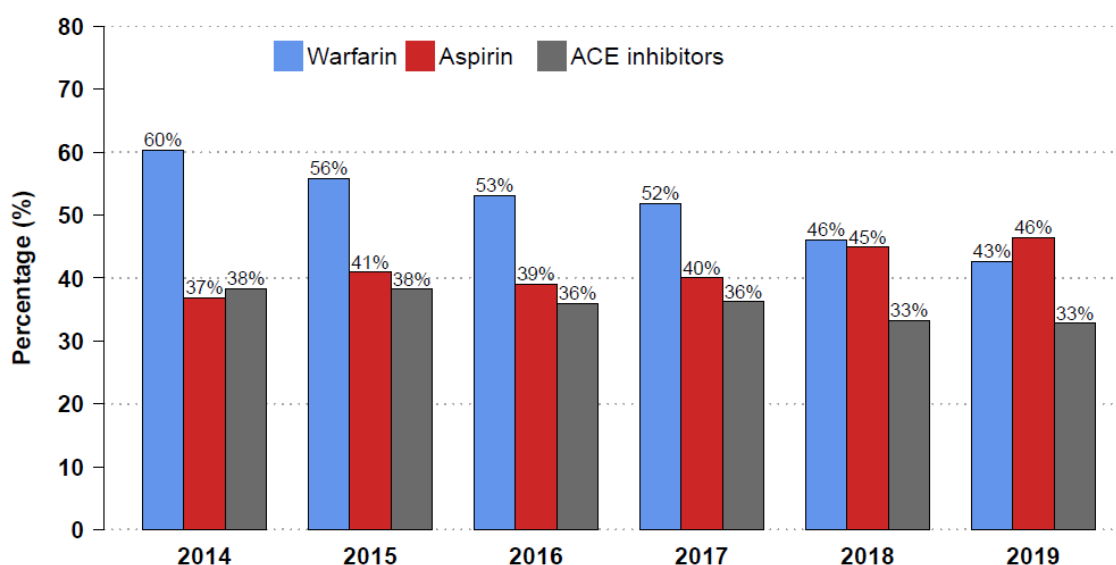
**Table 4 Post-Fontan anticoagulant medication usage at most recent follow-up (N = 1028)**

Anticoagulant medication	n (%)
Aspirin	498 (48.4)
Warfarin	466 (45.3)
Low Molecular Weight Heparin/Clexane/Enoxaparin	18 (1.8)
None	20 (1.9)
Not Mentioned	50 (4.9)

In addition to anticoagulant medications, a large range of other medications were mentioned in cardiology review letters. Among these other medications, at least one type of Angiotensin Converting Enzyme (ACE) inhibitor was used by 33.9% of participants (see Table 5) in 2018-2019. A comparison of Warfarin, Aspirin and ACE inhibitors over the last 5 years (2014-2019) is given in Figure 5.

**Table 5 Post-Fontan other medication usage at most recent follow-up (N = 1028)**

Other medication	n (%)
Lisinopril	178 (17.3)
Enalapril	103 (10.0)
Fruzemide	76 (7.4)
Sotalol	41 (4.0)
Digoxin	22 (2.1)
Perindopril	17 (1.7)
Ramipril	14 (1.4)
Captopril	9 (0.9)
Cilazapril	27 (2.6)



**Figure 5 Anticoagulation and ACE inhibitors 2014 -2018**

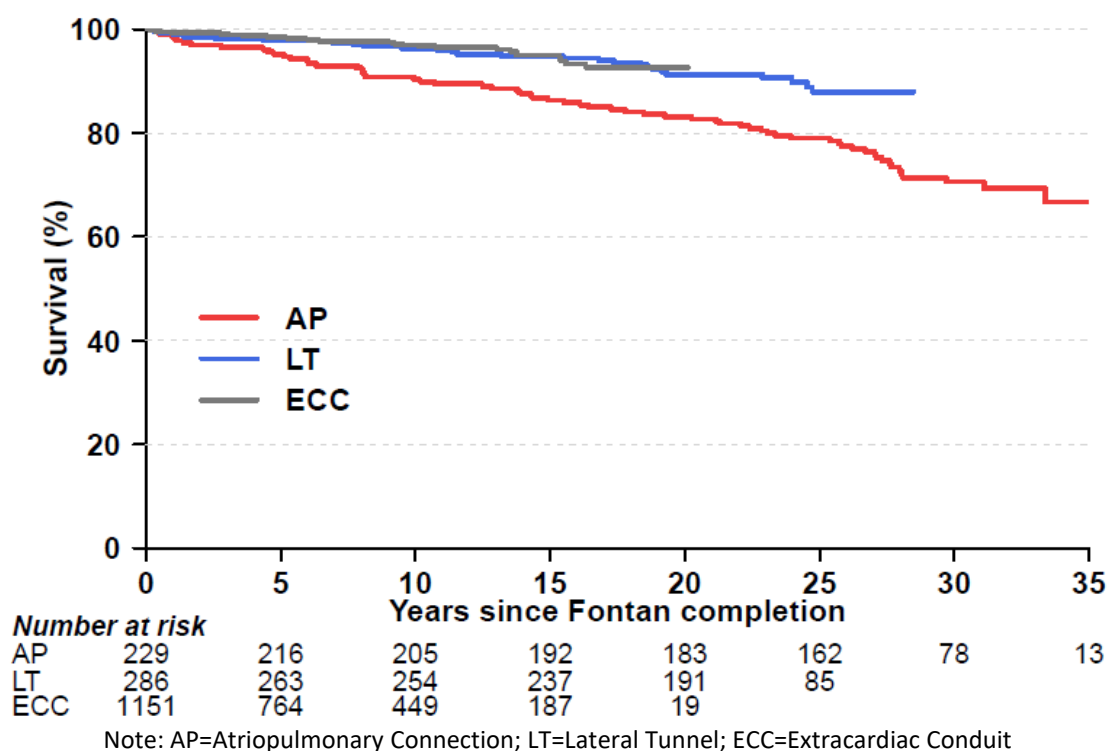


## SURVIVAL

The Registry enables us to describe the longest outcomes after Fontan surgery. The graph below describes the survival of those who was discharged from the hospital with a Fontan circulation. The start of the curve is therefore at 100% and decreases following the estimation of survival each year following the operation. These estimates are statistical calculations that are made as if we knew the outcomes of everybody, but they represent in reality the outcomes of those who have been operated many years ago. The number of patients under the bottom horizontal line indicates the number of patients on which these estimates are calculated. The longer the time, the less precise are the calculations.

For those who had an Extracardiac Conduit (ECC) procedure, the survivor rate at 10 years is 97%, 95% at 15 years and 93% at 20 years. For those who had a Lateral Tunnel (LT) procedure, the survivor rate at 10 years is 96%, 91% at 20 years and 88% at 27 years. For those who had the Atriopulmonary Connection (AP) procedure, the survivor rate at 10 years is 90%, 83% at 20 years, 71% at 30 years, and 67% at 35 years. To place this in context of the general population, a child at 5 years of age (the average age of the Fontan procedure) has a survivor rate of 98.5% of living another 35 years (Registry calculations on Australian Life Tables 2011-2013, Australian Bureau of Statistics, 2014).

This graph show us two important facts. The survival of those with a Fontan circulation is much better than previously thought. It was already known that the result of the two more recent versions of the Fontan operations (Lateral Tunnel and Extracardiac Conduit) was superior to the one of the original Fontan. Very importantly, it shows us that there is no sudden decrease in survival late after Fontan surgery. It was predicted by some that those with a Fontan circulation may deteriorate rapidly after an unknown number of years, but that does not stand true for a period as long as 30 years.



**Figure 6 Kaplan Meier Survival Curve by type of Fontan operation**

## IDENTIFYING RISK FACTORS AFFECTING SURVIVAL

Table 6 presents a summary of possible risk factors identified in Fontan Registry participants. The interpretation of a hazard ratio (HR) can be expressed as the risk or rate of an event increasing or decreasing with respect to the reference category (denoted by a HR=1). For example, in patients who had a BCPS (Bidirectional Cavopulmonary Shunt) operation, the rate of survival increases by (100% - 46% =) 54% compared to those without BCPS. For a continuous variable the hazard ratio (HR) represents the change in risk per unit increase of that variable. For example, an increase of 1 year in age at Fontan decreases the rate of survival by 5%.

**Note:** Not all the variables listed in Table 6 are risk factors for survival as this depends on the level of evidence as inferred by the last column “*p*”. If the value of *p* is small (usually a value of less than 0.05) then this corresponds to evidence that the variable affects the survival rate.

**Table 6 Risk Factors for Survival**

<i>Variable</i>	<i>Level</i>	<i>N</i>	<i>Deaths</i>	<i>HR (95% CI)</i>	<i>p</i>
Gender	Female	706	45	1	0.032
	Male	976	80	1.49 (1.03-2.15)	
Age at Fontan	Per year increase	1682	125	1.05 (1.02-1.08)	0.001
Fontan Procedure Type	Atriopulmonary Connection (AP)	229	65	1	<0.001
	Lateral Tunnel (LT)	286	26	0.45 (0.28-0.72)	
	Extracardiac Conduit (ECC)	1151	34	0.39 (0.24-0.62)	
Fontan Fenestrated	No	1021	91	1	0.96
	Yes	623	33	0.99 (0.65-1.50)	
Isomerism/Heterotaxy	None	1533	113	1	0.51
	Left Atrial Isomerism	47	5	1.68 (0.68-4.11)	
	Right Atrial Isomerism	68	5	1.12 (0.46-2.76)	
Cardiac Position	Normal	1528	112	1	0.54
	Dextrocardia/Mesocardia	154	13	1.19 (0.67-2.12)	
Predominant Ventricular Morphology	Left	943	75	1	0.16
	Right	567	42	1.45 (0.99-2.13)	
	Biventricular	101	5	0.92 (0.37-2.29)	
	Indeterminate	45	1	0.39 (0.05-2.83)	
Pre-Fontan Collaterals	No	865	62	1	0.65
	Yes	455	17	1.14 (0.65-1.98)	
Primary Diagnosis (morphological)	Tricuspid Atresia	359	33	1	0.05
	Double Inlet Left Ventricle	271	16	0.69 (0.38- 1.26)	
	Double Outlet Right Ventricle	233	20	1.12 (0.64- 1.97)	
	Atrioventricular Canal or AVSD	130	10	1.33 (0.65- 2.72)	
	Pulmonary Atresia with VSD	35	1	0.31 (0.04- 2.27)	
	Pulmonary Atresia with Intact	142	10	1.29 (0.63- 2.64)	
	HLHS	219	10	1.68 (0.80- 3.49)	
	Ebstein's Anomaly	16	4	4.03 (1.42-11.43)	
	ccTGA	109	7	0.90 (0.39- 2.03)	
	Other	154	14	1.39 (0.74- 2.60)	
Length of hospital stay	Per week increase	1294	83	1.08 (1.02-1.15)	0.007
	Per month increase			1.41 (1.10-1.81)	
Prolonged pleural effusion (post-op)*	No	1582	113	1	0.028
	Yes	100	12	1.93 (1.06-3.50)	
<b>Prior Surgeries</b>					
PA Band	No	1252	97	1	0.39
	Yes	391	26	0.83 (0.54-1.28)	
Norwood	No	1357	113	1	0.51
	Yes	286	10	1.25 (0.64-2.44)	

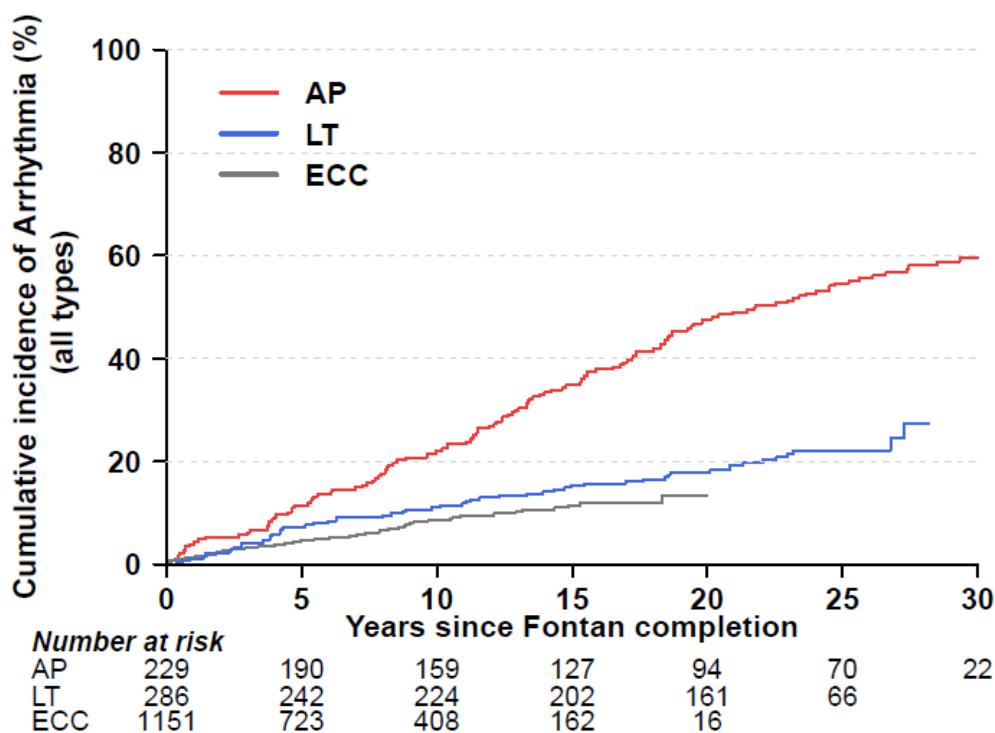
BT Shunt	No	1016	75	1	0.28
	Yes	627	48	0.82 (0.57-1.18)	
BCPS	No	682	96	1	0.001
	Yes	961	27	0.46 (0.29-0.72)	
BCPS with forward flow	No	1601	121	1	0.83
	Yes	42	2	0.86 (0.21-3.50)	
PA Reconstruction	No	1499	117	1	0.76
	Yes	144	6	0.88 (0.38-2.01)	
Aortic Arch Repair	No	1555	121	1	0.37
	Yes	88	2	0.53 (0.13-2.15)	
<i>Variable</i>					
Time-dependent variables	<i>Level</i>	<i>N</i>	<i>Deaths</i>	<i>HR (95% CI)</i>	
Protein Losing Enteropathy (PLE) or	No	1624	111	1	<0.001
	Yes	58	14	5.81 (3.28-10.3)	
Supra Ventricular Tachycardia (SVT)	No	1489	100	1	0.10
	Yes	133	22	1.55 (0.93-2.59)	
Arrhythmia**	No	1473	87	1	<0.001
	Yes	209	38	2.33 (1.49-3.65)	
Ventricular Dysfunction (moderate or	No	1531	88	1	<0.001
	Yes	151	36	7.31 (4.79-11.2)	

\*Defined as chest drains in-situ >30 days, or re-operations for effusions. \*\* Defined as SVT, flutter or fibrillation

**ARRYTHMIAS**

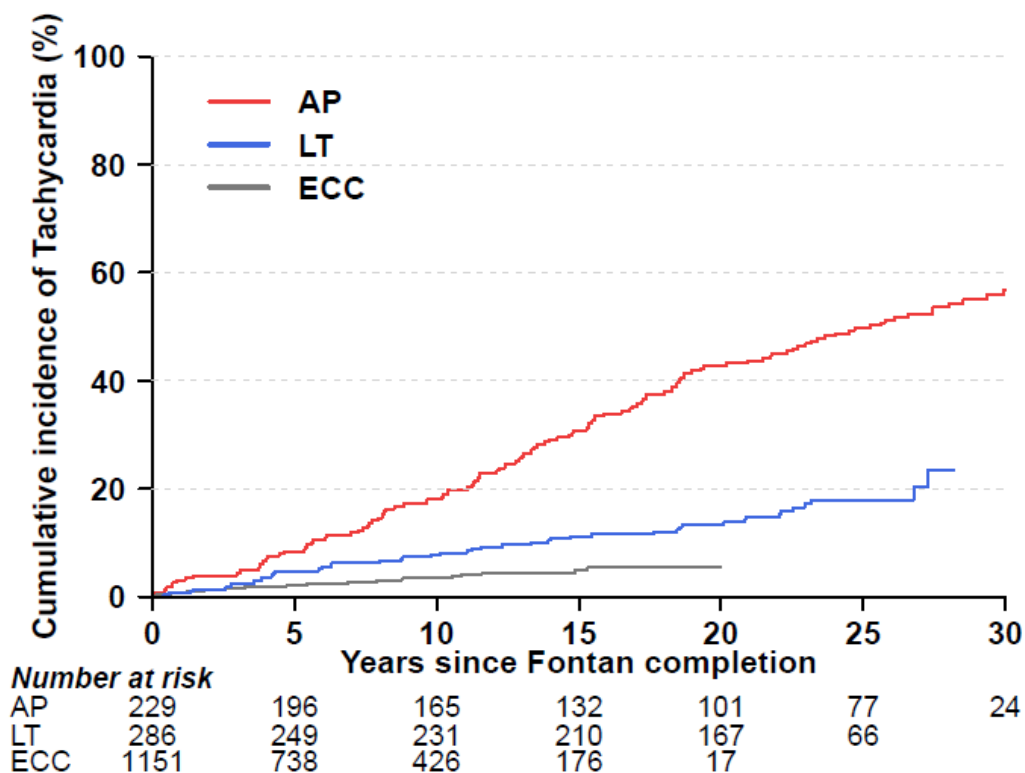
The Registry routinely collects information on the occurrence of arrhythmic events after Fontan surgery. The graphs below (Figure 7-9) describe the cumulative incidence of patients’ first arrhythmic event after Fontan surgery. The start of the curve is therefore at 0% and increases following the occurrence of arrhythmic events to estimate the cumulative incidence of arrhythmic events. Other features of the graphs are similar to that explained for the survival curve in Figure 6. The cumulative incidence of arrhythmic events is presented for 1) all arrhythmic events: flutter, supraventricular tachycardia, atrial fibrillation, sick sinus syndrome, bradycardia, junctional tachycardia, complete heart block and others (Figure 7); 2) tachycardia events: flutter, supraventricular tachycardia, atrial fibrillation and junctional tachycardia (Figure 8); and 3) bradycardia events: sick sinus syndrome, bradycardia and complete heart block (Figure 9).

For those who had an ECC procedure, the percentage who had an arrhythmia by 10 years (i.e., the cumulative incidence) post Fontan surgery was 9% and 13% at 15 years. For those who had a LT procedure, 11% had an arrhythmic event by 10 years and at 20 years it was 18%. Higher rates are seen in those who had the AP procedure, with 22% experiencing an arrhythmic event by 10 years and 48% at 20 years.



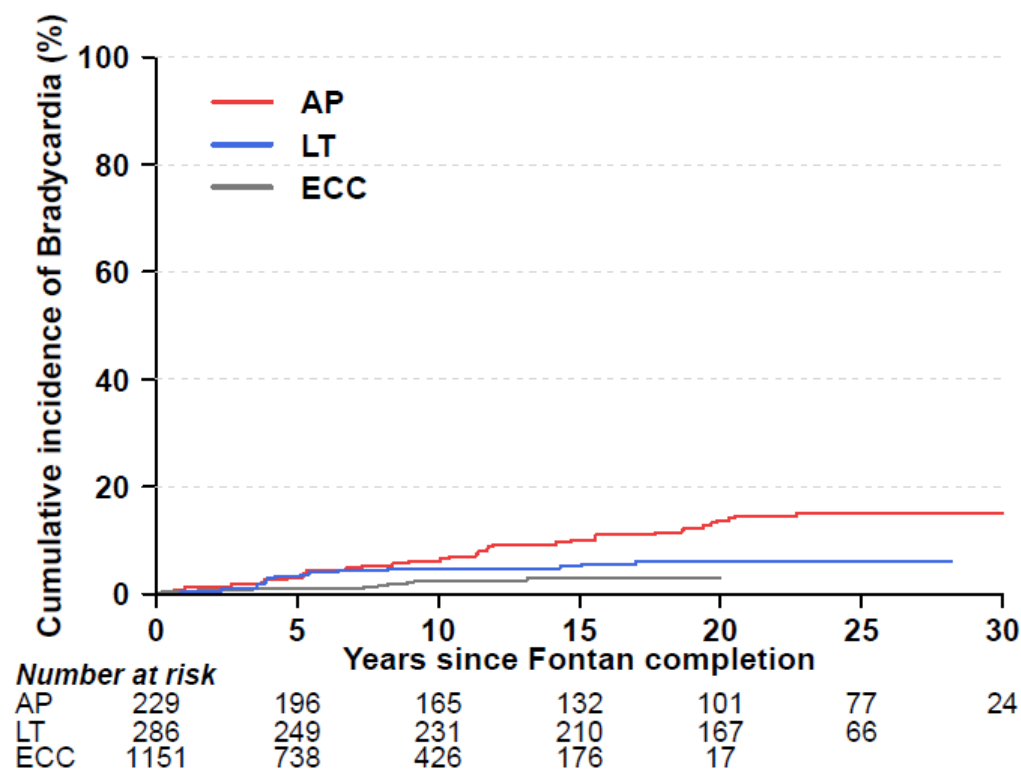
Note: AP=Atriopulmonary Connection; LT=Lateral Tunnel; ECC=Extracardiac Conduit

**Figure 7 Cumulative incidence of all arrhythmic event types by type of Fontan operation**



Note: AP=Atriopulmonary Connection; LT=Lateral Tunnel; ECC=Extracardiac Conduit

**Figure 8 Cumulative incidence of tachycardia by type of Fontan operation**



Note: AP=Atriopulmonary Connection; LT=Lateral Tunnel; ECC=Extracardiac Conduit

**Figure 9 Cumulative incidence of bradycardia by type of Fontan operation**

IDENTIFYING RISK FACTORS AFFECTING ARRHYTHMIA OCCURENCE

Table 7 presents a summary of possible risk factors for arrhythmias identified in Fontan Registry participants. Interpretation is as per explanation provided for Table 6.

**Table 7 Risk Factors for Arrhythmias**

Variable	Level	N	Deaths	HR (95% CI)	p
Gender	Female	706	115	1	0.23
	Male	976	158	1.16 (0.91-1.47)	
Age at Fontan	Per year increase	1682	273	1.05 (1.02-1.07)	<0.001
Fontan Procedure Type	Atriopulmonary Connection (AP)	229	134	1	<0.001
	Lateral Tunnel (LT)	286	58	0.33 (0.24-0.45)	
	Extracardiac Conduit (ECC)	1151	79	0.27 (0.20-0.36)	
Fontan Fenestrated	No	1021	193	1	0.14
	Yes	623	73	0.81 (0.62-1.07)	
Isomerism/Heterotaxy	None	1533	241	1	<0.001
	Left Atrial Isomerism	47	8	1.14 (0.56-2.31)	
	Right Atrial Isomerism	68	22	2.69 (1.73-4.16)	
Cardiac Position	Normal	1528	241	1	0.05
	Dextrocardia/Mesocardia	154	32	1.44 (1.00-2.09)	
Predominant Ventricular Morphology	Left	943	174	1	0.80
	Right	567	80	1.08 (0.82-1.40)	
	Biventricular	101	14	0.96 (0.56-1.66)	
	Indeterminate	45	5	0.70 (0.29-1.71)	
Pre-Fontan Collaterals	No	865	150	1	0.04
	Yes	455	32	0.67 (0.46-0.99)	
Primary Diagnosis (morphological)	Tricuspid Atresia	359	73	1	0.3
	Double Inlet Left Ventricle	271	48	0.89 (0.62-1.29)	
	Double Outlet Right Ventricle	233	48	1.15 (0.80-1.65)	
	Atrioventricular Canal or AVSD	130	20	1.04 (0.64-1.71)	
	Pulmonary Atresia with VSD	35	4	0.50 (0.18-1.37)	
	Pulmonary Atresia with Intact	142	13	0.63 (0.35-1.14)	
	HLHS	219	13	0.74 (0.40-1.34)	
	Ebstein's Anomaly	16	4	1.87 (0.68-5.12)	
	ccTGA	109	21	1.15 (0.71-1.87)	
	Other	154	29	1.20 (0.78-1.84)	
Length of hospital stay	Per week increase	1294	186	1.07 (1.02-1.12)	0.01
	Per month increase			1.32 (1.07-1.62)	
Prolonged pleural effusion (post-op)*	No	1582	259	1	0.86
	Yes	100	14	0.95 (0.56-1.63)	
<b>Prior Surgeries</b>					
PA Band	No	1252	207	1	0.38
	Yes	391	63	0.88 (0.66-1.17)	
Norwood	No	1357	250	1	0.60
	Yes	286	20	0.88 (0.55-1.40)	
BT Shunt	No	1016	150	1	0.52
	Yes	627	120	1.08 (0.85-1.38)	
BCPS	No	682	203	1	<0.001
	Yes	961	67	0.40 (0.30-0.52)	
BCPS with forward flow	No	1601	269	1	0.03
	Yes	42	1	0.15 (0.02-1.08)	
PA Reconstruction	No	1499	258	1	0.11
	Yes	144	12	0.63 (0.35-1.12)	
Aortic Arch Repair	No	1555	260	1	0.95
	Yes	88	10	1.02 (0.54-1.93)	

\*Defined as chest drains in-situ >30 days, or re-operations for effusions. \*\* Defined as SVT, flutter or fibrillation

## Publication List for 2019

The ANZ Fontan Registry continue to contribute important research to the Fontan community. A list of publications from 1<sup>st</sup> January – 31<sup>st</sup> December 2019 are given below:

### Atrioventricular Valve

- King G, Ayer J, Celermajer D, Zentner D, Justo R, Disney P, Zannino D, d'Udekem Y. [Atrioventricular Valve Failure in Fontan Palliation](#). *Journal of the American College of Cardiology*, 2019, Feb 26; 73(7): 810-822. doi: 10.1016/j.jacc.2018.12.025.PMID: 3078467

### Bone Health

- D'Ambrosio P, Tran D, Verrall CE, Attard C, Singh MF, Ayer J, d'Udekem Y, Twigg S, Celermajer DS, Cordina R. [Prevalence and risk factors for low bone density in adults with a Fontan circulation](#). *Congenital Heart Disease*, 2019, Nov; 14(6): 987-995. doi: 10.1111/chd.12836. Epub 2019 Aug 20.PMID: 31430042

### Exercise in Fontan

- Cordina R, d'Udekem Y. [Long-lasting benefits of exercise for those living with a Fontan circulation](#). *Current Opinion in Cardiology*, 2019, Jan; 34(1):79-86. doi: 10.1097/HCO.0000000000000582.PMID: 30444760
- Daley M, d'Udekem Y. In patients undergoing Fontan completion, does a younger age at operation result in better long-term exercise capacity and prognosis? *Interactive CardioVascular and Thoracic Surgery*, 2019, Feb 1; 28(2): 301-305. doi: 10.1093/icvts/ivy219.PMID: 30085097

### Liver and Renal

- Wilson TG, d'Udekem Y, Winlaw DS, Cordina RL, Ayer J, Gentles TL, Weintraub RG, Grigg LE, Cheung M, Cain TM, Rao P, Verrall C, Plessis KD, Rice K, Iyengar AJ. [Creatinine-based estimation of glomerular filtration rate in patients with a Fontan circulation](#). *Congenital Heart Disease*, 2019, May; 14(3):454-463. doi: 10.1111/chd.12746. Epub 2019 Jan 21.PMID: 30664330

### Long-term Outcomes

- Yong MS, Zhu MZL, du Plessis K, Weintraub RG, Hornung T, Winlaw D, Alphonso N, d'Udekem Y, Konstantinov IE. [Long-term Outcomes of the Fontan Operation in Patients With Total Anomalous Pulmonary Venous Drainage](#). *Annals of Thoracic Surgery*, 2019, Oct; 108(4): 1234-1241. doi: 10.1016/j.athoracsur.2019.04.073. Epub 2019 Jun 12.PMID: 31201782
- Marathe SP, Zannino D, Shi WY, du Plessis K, Kehr J, Perumal G, Sun J, Gentles TL, Ayer J, Sholler GF, Verrall CE, Alphonso N, Andrews D, d'Udekem Y, Winlaw DS. [Two Ventricles Are Not Better Than One in the Fontan Circulation: Equivalent Late Outcomes](#). *Annals of Thoracic Surgery*, 2019, Mar; 107(3): 852-859. doi: 10.1016/j.athoracsur.2018.08.024. Epub 2018 Oct 6.PMID: 30300642
- Shearer L, Justo RN, Marathe SP, Betts K, Venugopal P, Winlaw DS, Bullock A, Robertson T, Gentles TL, Celermajer D, d'Udekem Y, Alphonso N. [Augmentation of the pulmonary arteries at or prior to the Fontan procedure is not associated with worse long-term outcomes: a propensity-matched analysis from the Australia-New Zealand Fontan Registry](#). *European Journal of Cardiothoracic Surgery*, 2019, May 1; 55(5): 829-836. doi: 10.1093/ejcts/ezy376.PMID: 30496386

### Long-term Outcomes *continued*

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