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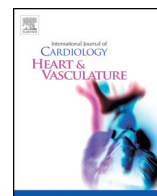
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“Will she live a long happy life?” Parents' concerns for their children with Fontan circulation

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ABSTRACT

Background: Families of children at the worst end of the congenital heart disease endure a significant burden which is often not clearly delineated in the clinical literature. We examined the greatest concerns of parents whose children have a Fontan circulation.

Methods: Parents ($N = 107$) of children in the Australian and New Zealand Fontan Registry completed online surveys with open-ended and closed questions. A qualitative method approach incorporating thematic analyses was used.

Results: The greatest concerns for parents of a child with a Fontan circulation were centered on fear of death for their child and psychosocial well-being, followed by lesser themes around anti-coagulation use, pregnancy and financial burdens.

Conclusions: Fear of death and the psychological well-being of their children were the main parental concerns. It highlights the need to clearly communicate information on outcomes to families, and the need for family-focused psychological interventions to improve the psychosocial functioning of both parents and young people.

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1. Introduction

The Fontan procedure is the last in a series of operations for babies born with a single ventricle physiology. In the last decade, it has become clear that survival outcomes were better than expected, with the hope that up to 80% of those who have undergone the procedure surviving 30 years after the procedure [1–4]. This population faces a considerable burden of disease. Half of them are expected to face a major complication requiring hospitalization before adulthood, they are known to have lim-

ited exercise capacity, and they face multi-systemic complications related to elevated central venous pressures [5,6]. This extraordinary journey through the health system generates, without doubt, anxieties and, at times, extreme difficulties for the families of these children, but these pressures have not been well investigated.

Parents of complex congenital heart disease children experience elevated stress in comparison to the general population, and this is associated with adverse outcomes, including decreased physical and psychological wellbeing in both children and parents [7]. Mental health risks (particularly in the period following cardiac surgery) include anxiety, depression, high levels of distress, trauma, and symptoms consistent with PTSD [8]. Extensive financial, familial and emotional costs have also been identified in the literature of CHD parents, and this frequently co-present with uncertainty and distress which are linked to the complexity of the child's disease and parents' socioeconomic status [9,10].

We have created a bi-national Registry in Australia and New Zealand and work closely with patients and their families [11,12]. This interaction resulted in the creation of an advocacy group, the Australian and

Abbreviations: AVSD, Atrioventricular Septal Defect; ANZFAC, Australian and New Zealand Fontan Advisory Committee; ccTGA, Congenitally Corrected Transposition of the Great Arteries; DILV, Double Inlet Left Ventricle; DORV, Double Outlet Right Ventricle; HLHS, Hypoplastic Left Heart Syndrome; INR, International Normalized Ratio; PLE, Protein Losing Enteropathy; PTSD, Post-Traumatic Stress Disorder (PTSD).

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New Zealand Fontan Advisory Committee (ANZFAC). Their first task was to survey the greatest concerns of parents of this population.

2. Methods

Ethics approval was obtained through the Royal Children's Hospital, Melbourne, Australia. An anonymous online survey assessed the needs of parents through the Australian and New Zealand Fontan Registry in 2015. In addition to basic demographics, this purpose built survey used both open-ended and closed questions (Table 1).

2.1. Analyses

The major focus of this paper is around parents' greatest self-reported concerns for their child with a Fontan circulation. We used thematic analyses to showcase the rich and complex stories of parent experiences of their child's (patient's) health care journey [13]. Survey responses were analyzed by the Fontan Registry Research Coordinator and Fontan Registry Steering Committee Chair/Founder, who both work closely with the Fontan community, the Registry, and its associated research projects. Similar to Braun and Clarke, we viewed thematic analysis as an "essentialist or realist method which reports experiences, meanings and the reality of participants" [14]. A theme was construed as a pattern found in qualitative information that describes and organizes the information [15]. The manual coding process followed a three-step progression, including (a) developing concepts and categories to organize data into a framework of ideas, (b) comparing data instances, cases and categories for similarities and differences, and (c) unifying key themes [15]. This was repeated until a point of saturation in terms of conceptual depth was reached in the themes [16], in addition to which the key themes were also checked for resonance and validity with ANZFAC (co-authors) as representatives of the parent and patient community. Where relevant, descriptive quantitative information from the rest of the survey are referenced under key themes identified.

3. Results

A total of 107 parents of children with a Fontan circulation participated in the survey from 338 Fontan Registry parents with e-mail addresses (response rate 32%). The geographical spread of participants was similar to that in the broader Australian and New Zealand Fontan Registry. The majority of parents were in married/de facto relationships ($n = 84$, 80%), and all were individual respondents, bar one couple. The majority of children (represented by parents) were male ($n = 68$, 64%), the average age was 11 years ($SD = 5$), and the average age at time of Fontan completion 4 years ($SD = 2$). No data around racial/ethnic identities, parents' gender or level of education were gathered. Major diagnoses included: Hypoplastic Left Heart Syndrome (HLHS) ($n = 39$, 36%), Tricuspid Atresia ($n = 38$, 36%), Congenitally Corrected Transposition of the Great Arteries (ccTGA) ($n = 16$, 15%), Atrioventricular Septal Defect (AVSD) ($n = 35$, 33%), Double Outlet Right Ventricle (DORV) ($n = 37$, 35%), Double Inlet Left Ventricle (DILV) ($n = 3$, 5%), and Pulmonary Atresia ($n = 4$, 7%). These diagnoses reflect an overrepresentation of participants with the more severe conditions (i.e., HLHS) in the Fontan Registry. Almost half of the parents did not know what type of Fontan operation their child had ($n = 49$, 46%), with the remaining majority reporting that their child had an extra cardiac conduit ($n = 38$, 36%).

Most of the parents who had completed the survey answered this question ($n = 93$, 87%), and responses varied from one word to several sentences per participant, many touching on several key themes in their answers. The two major themes are described below (Fig. 1).

3.1. Theme 1: fear of death/uncertainty around life expectancy

Fear of death was the greatest concern for most parents coloring the manner in which they considered their child's physical health and treatment. It was pervasive, omnipresent, and a dark burden on parents. In general, this theme was more closely related to medical/physical components, rather than socio-emotional aspects. Responses were often summarized with succinct responses to the question around 'greatest concerns' e.g., "Death" (Participant 24), whereas others were

Table 1
Survey questions.

Sample questions	Response options
Your greatest concern(s) for your child with a Fontan circulation is...	Open-ended
In regards to anti-coagulation, what are your greatest concerns/issues?	Open-ended
How many times a week does your child exercise?	Nil; Once a week; Twice a week; Three times a week; Four times a week; Five times a week
What type of exercise?	Open ended
How would you rate your child's capabilities, in comparison to their peers, in each of the categories below?	Significantly less; Slightly less; About the same; Slightly more; Significantly more
Physical activity/exercise tolerance	
Emotional wellbeing	
Behavior	
Education & learning	
How frequently does your child experience any of the following symptoms?	Always; Sometimes; Occasionally; Never; Do not know; Already present prior to Fontan
Leg pains/leg muscle cramping/leg irritability	
Migraines (prolonged headache)	
Mouth ulcers	
Difficulty sleeping	
Night terrors	
Does your child seem sad or display signs of depression	
Nail biting	
Does your child have learning or concentration difficulties	
Does your child seem to get angry or lose his/her temper easily?	
To what extent do you feel your child's school support his/her needs	Not at all; Moderately supportive; Very supportive (sliding scale)
Have you accessed psychological/psychiatric/counselling services previously in relation to your child's health?	No; Yes (please describe your experience of accessing these services below)
Would you be interested in accessing psychological/psychiatric/counselling services?	Yes; No; Not sure
Has your child's medical journey impacted you or your family financially?	No; Yes (please specify how this has impacted below, e.g., employment opportunities, financial hardship)

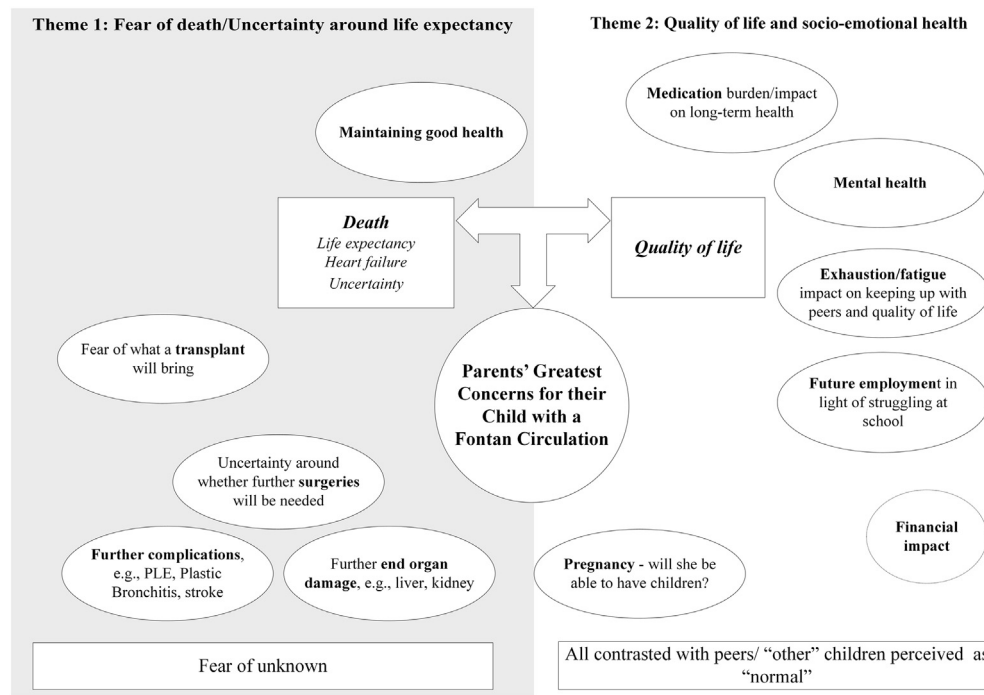


Fig. 1. Key themes of parents' greatest concerns for their child with a Fontan circulation.

less direct, e.g., “Life expectancy” (Participant 21), and “How long will his heart be able to cope with the Fontan circulation before he either needs a heart transplant or dies!” (Participant 72).

Fear of death was compounded by uncertainties, e.g., “What other problems can occur from having a Fontan?” (Participant 87). In contrast with these uncertainties, some parents were clear on what they perceived were linked to a deteriorating Fontan circulation, including the impact on end-organs/Fontan-related complications. This linked to their fear of complications and eventual death for their child, e.g., “Ventricular fibrillation leading to heart failure, or Fontan circulation failing over time. Trauma to heart through physical contact/accident, or infection” (Participant 47). Additional concerns linked to Theme 1 included: The effect on other organs (e.g., “How her body will cope with her Fontan in the future. How will her heart hold up and her other organs”, Participant 7); complications (including Protein Losing Enteropathy (PLE), Plastic Bronchitis and heart failure) (e.g., “That he will develop PLE, plastic bronchitis or other potential complications that could be life limiting”, Participant 82); transplantation (e.g., “That his heart will fail and lead to transplant”, Participant 38); further surgeries (e.g., “Should I expect her to need any more surgery down the track?”, Participant 61); and maintaining good health (e.g., “The longevity of the procedure. Will it be enough to help him through his adult life?”, Participant 14).

3.2. Theme 2: quality of life and socio-emotional health

Quality of life connected with Theme 1: Fear of death/Uncertainty around life expectancy, and was often phrased together by parents (e.g., “longevity, quality of life”, Participants 10, 42, 26, 66, 74, 83, 84, 91). Parents want their children with a Fontan circulation to have a long life, but also a high quality ‘happy’ life, e.g., “Will she live a long happy life?” (Participant 31).

Quality of life also stood by itself as a greatest concern for some parents. The Quality of life/Socio-emotional health theme as a whole was contextualized to what is perceived as ‘normal’, e.g., “Will she be able to lead a ‘normal’ life, have children?” (Participant 55).

What is perceived as ‘normal’ was positioned in relation to peers, and around developing ‘normal relationships’, e.g., “That life for her is a never-ending physical challenge that has affected her view of relationships and the world in general. I fear she will never realize what it feels like to be truly OK” (Participant 107).

3.2.1. Medications and anticoagulation

‘Medications’ triggered concerns for its physical impact and socio-emotional burden. In terms of anticoagulants, over two thirds of the parents surveyed indicated that their child was on warfarin ($n = 68$, 64%), and one third on aspirin. Over three quarters of all the parents whose children were on warfarin ($n = 53$, 78%) had concerns. These included: Excess/internal bleeding (e.g., “Possible hemorrhage due to combinations with other medications and potential trauma from an incident (especially as he is a 14-year-old- boy!)” (Participant 60); bruising (e.g., “All the side effects of warfarin - knocks/bruises are a concern, limited in sports.”) (Participant 75); risk of stroke (e.g., “We want it to be strong enough that he doesn’t have a stroke due to blockage”) (Participant 80); low bone mineral density (e.g., “The stability of his INR and links to long term use causing osteoporosis.”) (Participant 105); the ‘unknown’ effect of long term use on the body (e.g., “The added risks to her health and wellbeing by taking this medication long term? Is it really safe enough to take long term? What are the alternatives if not safe though for long term use?”) (Participant 31); dosage - difficulties with keeping INR in range, including as a result of diet, season, adolescence (e.g., “Keeping the medication at the right level. It’s a balancing act and can be affected by so many things”) (Participant 55).

Parents who were using home International Normalized Ratio (INR) monitoring reported this being a much needed help for families, e.g., “Has just improved greatly with our own machine at home and managed by hospital. Made a huge difference to time and efficiency” (Participant 63).

Parents whose children were on aspirin had concerns around excessive bleeding, and the long term impact on organs, specifically, liver, kidneys and stomach. A number of the children had recently been moved to aspirin, and overall parents expressed relief. Some

Table 2
Parents' self-report of mental health/psychological symptoms in their children with a Fontan circulation.

	Always	Sometimes	Occasionally	Never	Do not know	Already present prior to Fontan
Difficulty sleeping (n = 106)	16%	21%	28%	35%	0	7%
Night terrors (n = 106)	5%	15%	23%	55%	2%	3%
High levels of anxiety (n = 105)	15%	32%	30%	21%	2%	4%
Sadness of signs of depression (n = 105)	2%	19%	27%	49%	3%	1%
Learning or concentration difficulties (n = 106)	26%	23%	23%	27%	1%	4%
Anger or losing temper easily (n = 106)	20%	39%	20%	22%	1%	3%

questions however, remained, including: "Aspirin v warfarin...which is best???" (Participant 84).

3.2.2. Mental health

Parents noted 'Mental Health' as one of their greatest concerns, e.g., "She has been diagnosed with post-traumatic stress – dealing with health worries" (Participant 39).

Separate to the question of greatest concerns, parents also indicated (summarized in Table 2) whether their child experienced symptoms which might be indicative of mental health or psychological issues.

Additional symptoms/behavioral issues reported were nausea, stomach pains, bed wetting, and forgetfulness. Two further key themes stood out: fatigue and anxiety, with fatigue often occurring as a result of overexertion (see Exhaustion/Fatigue below). With regards to anxiety/stress/not being able to cope, and the need to self-soothe, of particular concern was the young child who at age 6 had started mentioning suicide, e.g., "Started to chew on his clothes... Talks of killing himself every now and then (started age 6) 2 years post Fontan" (Participant 78).

3.2.2.1. Accessing psychological services/other support. Almost half of the participants (44%) had accessed psychological/psychiatric/counselling support, and 52% indicated that they would be interested in accessing this type of support. Most parents rated schools as moderately to very supportive of their child's needs.

3.2.3. Exhaustion/fatigue

A number of parents raised fatigue and exhaustion as among their greatest concerns for their child living with a Fontan circulation, e.g., "[He] has progressively become more inactive, fatigued and unable to attend school full time" (Participant 60). For others this was marked in relation to peers, e.g., "Ability to function like others his age. Exhaustion is a major factor" (Participant 22).

3.2.3.1. Self-reported exercise. With regards to exercise, 46% were exercising less than three times per week, including walking, running, gym, team sports, swimming, cycling, dancing, basketball, tennis and playing with siblings. Most parents (88%) rated their child as slightly/significantly less, capable than their peers in physical activity/exercise tolerance.

3.2.4. Future employment

Parents worried about the future employment abilities of their child, particularly in the context of missing school as a result of poor health e.g., "We are concerned for his future as to how more debilitating things may become for him, and how this will impact on his education and future work opportunities" (Participant 60).

3.2.5. Pregnancy

Parents had concerns around how the Fontan circulation will impact their daughters' ability to have children, e.g., "Will she be able to have children and if so how safe will this be for her?" (Participant 31).

3.2.6. Financial impact

Two thirds (66%, n = 69) of parents reported that they were financially impacted by their child's medical journey. While this was not mentioned in any responses around *greatest concern*, the impact of the answers on its own, warrant inclusion here. It should also be prefaced by acknowledging that parents' recognition of financial impact does not imply they begrudge their child with a Fontan circulation, and a number of parents expressed this type of sentiment, e.g., "He's worth every penny and the time we have spent with him is priceless" (Participant 29).

A number of key themes are highlighted around financial impact, including the flow-on effect on employment/income generation, travel costs and expenditures associated with prolonged hospital stays, additional medical expenses, difficulties with planning financial future

Table 3
Financial impact and its sub-themes.

Quotations	Sub-theme	Major theme
"Quit our jobs and moved in with mother in law for first year of her life." (Participant 88)	Unable to work during hospital stays	Employment
"After 10½ years we are finally in a place where we can consider having two working parents, rather than one due to the time he has off school and the number of appointments we attend. This has cost us (a conservative estimate) \$320,000 in lost income." (Participant 29)	One parent not able to work/reduced hours/difficulties with caregiving support	
"I have chosen part time over full time work, and a lesser paid job than I am qualified for – to access appointments and to be able to attend any health issues as needed." (Participant 42)	Taking on more flexible working arrangements	
"I did not return to work after his birth, preferring to wait until after his Fontan surgery and he started school." (Participant 97)	Not being able to return to work for an extended period/ongoing	
"Huge financial strain, car parking at the hospital, food/toiletries whilst staying in hospital for long periods, everyone forgets that you still have your 'normal' expenditures e.g., rent/mortgage, bills, insurance petrol etc." (Participant 91)	Travel costs for those traveling	Travel costs
"We have had to move closer to services and take on a bigger mortgage" (Participant 27)	Moving closer to treating hospital	
"Ongoing costs with medications, INR strips. We have experienced numerous other health needs e.g., podiatry, extensive dental, optometry, speech, ENT [otolaryngologist], asthma specialists." (Participant 42)	Additional medical services; Costs ongoing into adulthood	Additional medical expenses and ongoing financial costs
"After a 3+ month stint at Hospital my husband's business operated at a loss as neither of us worked during this time. We spent all our savings, investments, and were one mortgage payment away from losing our home." (Participant 72)		Financial hardship/difficulty planning for financial future

on reduced incomes, financial hardship for some, and ongoing financial costs into adulthood (summarized in Table 3).

4. Discussion

The findings are based on a survey that was designed as a parent and adult patient initiative with little involvement of medical specialists. While limited to self-report it presents a snapshot of the voice of parents and their greatest concerns for their child with a Fontan circulation. As is common in survey data, the parents who participated were likely more motivated to voice their concerns, and therefore caution should be applied to generalize these findings to all parents in the Fontan community. It was not designed as a psychological or socio-economic study. Nonetheless it reveals findings highly relevant to our understanding of our CHD patients and their families.

The most important and constant concern for parents of children living with a Fontan circulation, as is also common in other chronic diseases [19], was the fear of losing their child. Fear of death can evoke powerlessness, separation and meaninglessness, which, when it comes into conscious awareness, often triggers defenses to remove these thoughts (i.e., through suppression/denial), but which might also include strategies such as maintaining the optimum health of themselves and their loved ones [20]. Fear of death points to a heightened anxiety state often presenting comorbid with psychopathologies (i.e., anxiety and depression disorders) [20]. Parents of children with critical congenital heart defects are at high risk for mental health morbidity, including over 80% presenting with clinical symptoms of trauma, 25–50% presenting with depression and/or anxiety, and up to 30% developing PTSD [8]. Mental health risks are highest in the weeks to months following cardiac surgery, and severity of mental health symptoms are positively correlated with severity of heart defect [8]. The building of the Australian and New Zealand Fontan Registry led to the creation of a close-knit community, informed by regular e-mails, newsletter, a yearly education day, and an active Facebook page. Informal feedback received lead us to believe that providing more accurate information on outcomes is better than being left in the dark. We hope that future generations of families supporting a child with a Fontan circulation will have decreased anxiety if they are better informed, giving them, like with other conditions such as cystic fibrosis, a sense of control over their condition (19). Previous research identify interventions which focus on striving to change families' illness perceptions and coping mechanisms, as most effective in reducing parental stress [21]. Future research could explore the efficacy of newer methods using social media or e-health resources [22,23]. Given that parenting stress has also been linked to family resources [7], these findings highlight the important role which community support organizations could play in providing social and financial support to these families.

Parents report at least mild psychological/mental health issues in their children. This concern seems to be more important to them than their child's physical capacity. Half of these families had sought support. This finding is of interest because most of our efforts focus on obtaining the best cardio-respiratory fitness for our patients, and only a small proportion of health expenses in Australia and New Zealand are directed towards psychological services. Early psychological intervention integrated into primary care settings are cost-effective and efficacious in treating mental health problems in adult cardiac care settings [24,25], with similar integrated pediatric-based services showing promising results in improving mental health outcomes for parents and children [26,27].

The burden of being on warfarin was evident with three quarters of parents having concerns. In a period of equipoise about the advantages of warfarin over aspirin, we should realize that our decision to opt for warfarin is accompanied by a clear burden for the families [28–32].

Despite the fact that two thirds of these families were financially affected, the financial burden of the condition was not quoted as a burden to any of these families. It is possible that financial burdens

might be underreported in this sample given the survey method using email and its potential link to higher socioeconomic status, thus care should be taken to generalize this to all parents of a child with a Fontan circulation. Nonetheless, the commitment of these families and their sacrifice for their children call for our admiration.

Where previous studies have focused on the critical period following cardiac surgery or in parents of young children [7,33], this study highlights that parental concerns continue well into adolescence and early adulthood, and it adds to our knowledge on the long-term concerns for parents of children with serious congenital heart disease. Future research exploring the trajectory of parental concerns in the context of mental health and coping, would further add to our understanding of stress-points for families, and where further interventions, in addition to early intervention [24,25], might prove most beneficial.

Qualitative research on patient perspectives is not sufficiently present in our current literature [17], in particular for quality of life of young people with a Fontan who will soon be moving adulthood [18]. Thus, qualitative investigation of patients and family satisfaction is a mandatory step to improve the provision of care. This current study calls for better information provision on outcomes to our patients, a greater understanding in healthcare professionals of the psychosocial challenges that families face [34], and for investment in early psychological assessment and support for parents and children with a Fontan circulation.

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Disclosures

Prof Yves d'Udekem is a consultant for the companies MSD and Actelion. The remaining authors report no relationships that could be construed as a conflict of interest.

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