



Flipping the script: The power of positivity in Fontan care

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ABSTRACT

Clinicians' words have a profound impact on a patient's attitude and outlook on life. Positive language, which acknowledges the uniqueness of each patient and highlights personal triumphs, can relieve anxiety and empower individuals to pursue their passions. Not only does this enhance emotional health, but an increasing body of literature indicates that positivity and mindful awareness improve aspects of physical health critical to Fontan optimization as well. There are still many unknown factors influencing long-term outcomes and quality of life. Thus, by acknowledging that today's "best available evidence" may not be "truth" over time for an individual, clinicians may open up possibilities for patients to define their own meaningful lives. Through small shifts in language, clinicians may change the narrative to the positive for individuals with Fontan circulation and in doing so offer patients the gift of "Yes" to the fullness of life.

1. Introduction

*Yes is a world
& in this world of
yes live
(skillfully curled)
all worlds*

e.e. cummings

This special edition highlights advances in Fontan care, including bioengineering to optimize flow dynamics, the use of MRI for risk stratification, exercise as prevention and treatment, and the national Fontan Outcomes Network. Such advances in research and outcomes have set the stage for a paradigm change in Fontan care- from awaiting failing physiology and death to anticipatory optimization of long-term outcomes. Along with this paradigm shift, it's time to "flip the script" - to change our narrative to the positive for people with Fontan circulation.

Language matters. Perspective matters. As clinicians and patients, we have extraordinary opportunities to listen and to bear witness. Lives get expressed in vibrant detail by unique voices, and worlds come alive before us. It is important to remember that these stories are still unfolding. What is "best available evidence" today may not be "truth" over time for an individual or the field. Moreover, the meaning of this moment or this lifetime may not be clear yet for any of us. From this front row seat on mystery, we are offered a remarkable invitation to use our words for empowerment. Our "yes" may be a world in which all worlds live.

Consider this "Yes" which opened a world for one of us- who is thriving as a newlywed, medical student, and individual with Fontan circulation.

"My surgeon told my parents, 'Taylor may never be an Olympian, but she can do whatever she wants.' So after my surgeries, my parents signed me up for a non-Olympic activity, ballet. Through dance, I was able to learn a lot about my heart and how it responded to physical exertion. My cardiac team empowered me to enroll in a rigorous studio and taught me how to best fuel my body, so I could keep up with the other dancers. I was always reminded to listen to how I felt and reassured if any unique symptoms occurred. My doctors gave me the gift of confidence which allowed me to pursue what I loved to do." (Figs. 1 and 2)

Focusing on positive language requires only a small shift. Asking individuals with Fontan circulation what they are most proud of, excited about, or aspire to be allows them to identify what is most meaningful to them. It offers a respectful yes- at every stage, your unique gifts and fullest expression of yourself will define you, not your diagnosis or limitations. Encouraging patients to recognize their own strengths- rather than defining "normal" and focusing on areas where they fall short- boosts confidence and self-esteem and enables them to celebrate when they surpass their personal goals and expectations. One simple approach emphasizes individuality while mitigating potential feelings of isolation: "Yes, you are unique, but you are not alone. Would you like to see pictures of kids and adults with your condition or go to a

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Fig. 1. Author, Taylor Houlihan, as a junior in high school, performing the role of a demi-soloist in the Waltz of the Flowers during her local studio's Nutcracker performance.



Fig. 2. Taylor Houlihan, now a second year medical student, and her husband Nathan, a third year medical student, celebrating her white coat ceremony at Sidney Kimmel Medical College in Lubert Plaza.

fun camp with them?" Try showing a picture from the internet of a thriving adult who publicly shares about life with a Fontan (like us) or hand out a heart camp brochure (Figs. 3 and 4). Similarly, asking a question typical in other childhood settings, "What do you want to be when you grow up?" is a powerful affirmation to patients and parents-you, the clinician, believe the child is going to grow up.

Seeing positive role models is key for all youth and perhaps even more for those with congenital heart disease. Since now over half of all individuals with Fontan physiology are over 18 years old, it is not hard to find us and share inspiration.

"My name is Alicia Wilmoth. I revel in life. I am happily married, am a proud mom of 3 amazing children- twin 6-year-olds and an energetic 2-year-old- and practice as a pediatric cardiac physician assistant. My life has always been full of friends, laughter, and a positive perspective. For my 40th birthday, I made it a point to have a



Fig. 3. Taylor Houlihan, as a junior at Wheaton College, performing her own choreographed piece in the annual fall show hosted by the campus dance group, Zoe's Feet.



Fig. 4. Author, Alicia Wilmoth, rock climbing and completing high ropes courses at Adult Congenital Heart Camp organized by Cincinnati Children's Hospital Medical Center.

big celebration and we flew to Las Vegas. In my career, I have had the honor to work in the best institutions in my field. Living with a single ventricle, I have undergone a Blalock-Taussig-Thomas shunt, a Glenn, and a Fontan, and as a clinician, I have been a surgical first assistant in each of those operations. I am inspired by people with the desire, determination, and willingness to focus on finding life-saving solutions. I recognize that there are so many things that we don't know. However, I prefer that we not assume going through these experiences only brings trials- it brings many, many, triumphs too!" (Figs. 5 and 6)

Clinicians' words carry tremendous weight and may have the power to shape a patient's attitude. They are often replayed in patients' minds and repeated to family and friends. Consider the cumulative impact over decades of reiteration of "Yes, we are doing everything we can to preserve his or her heart function and to build up strong muscles and bones, and yes people all over the world are actively researching novel treatments like exercise, pumps, and stem cells," compared to "We will check every year for heart failure and failure to thrive, but Fontan failure and heart transplant are inevitable." There is increasing



Fig. 5. Alicia Wilmoth with her husband, Kenny, celebrating her 40th birthday in Las Vegas.



Fig. 6. Alicia Wilmoth assisting in open heart surgery with Dr. Roosevelt Bryant at Cincinnati Children's Hospital Medical Center.

evidence that a patient's positive attitude affects cardiovascular status [1–3]. In other chronic disease populations, mindful awareness has been linked to improved outcomes in many domains key for Fontan outcomes- lower heart rate and blood pressure, better endothelial function, decreased inflammation, less anxiety, improved executive function and memory, better immune function, epigenetic alterations of chromosomes, and improved telomere length [4–12]. Additionally, in teens and young adults, social connection and self-management correlate to improved outcomes [13,14]. The “power of one” - having one adult who believes in their potential- has been shown to be instrumental in children with early adverse childhood experiences [15–17]. In adults, strong social supports and an appreciation of joy are positive predictors of physical health and happiness over decades [18,19]. Conversely, levels of patient and parent anxiety are correlated with

worse health-related quality of life in Fontan patients [20,21]. Negative language may create invisible barriers to patients flourishing, cause them to internalize “sick patient status,” or prevent them from pursuing their dreams, including marriage or career.

“Words to me are the most pivotal part of my care. When I hear ‘Failing Fontan’, I perceive it as saying I am failing. You just read my bio- who considers that failing? Truth is, I have never failed anything in my life. I’m a thriver. I have always fought; even the battles I don’t win, I fight. Support me in this. Simply speaking words of positivity into existence not only prevents my mind from going down rabbit holes but encourages me to keep fighting the good fight. I’ve been dealing with so much- I don’t need to add words to that list.” – AW (Fig. 7)

Of course, transparency is important. Clinicians must warn of potential significant potholes or hairpin turns on the roadmap, as well as discourage behaviors that might cause physical damage. However, caution and caring for oneself can be a double-edged sword. This can lead individuals with Fontan circulation to live in bondage to fear, believing the lie that their condition prevents them from living life to the fullest. Every patient deserves an individualized approach; a Fontan diagnosis should not instantly imply they will experience certain complications [22]. If and when limitations are encountered, solutions may be posed in a positive framework, with the shared goal of maximizing opportunity as much as possible.

“Sometimes I am shocked to read research articles about the Fontan circulation. I think, ‘They can’t possibly be talking about me.’ I don’t have failing organs or a neurocognitive deficit. I’m thriving in medical school and exercise almost every day. Although I realize this is not everyone’s story, it worries me that this is the impression new parents receive when their child is diagnosed with a congenital heart difference. I have certainly had difficulties along the way, but I have never taken ‘No’ as an answer and never let the fact that my heart works differently change my goals in life. I want families to understand this is not a tragedy but simply an obstacle, and obstacles are common to the human experience.” – TH (Fig. 8)

Our primary aim is to do no harm. Given that we don’t yet understand how to predict the trajectory for an individual Fontan patient, conventional language may be giving false despair, in an attempt to avoid unfulfilled promises. Instead, “we can only offer the best we know and what that means remains to be seen” leaves something very important for people. We will accompany each other as fellow travelers in that process of revelation. In time we are going to know more and more, and when we do, we’ll be there with you. Accompaniment is a gift and eases fear. “We don’t know yet” is not abandonment. It is welcoming life into the exam room and acknowledging that there are dimensions of life that are beyond science.

As an example, consider the life of famed cardiovascular pioneer, Dr. C. Walton Lillihei. As a senior resident, at age 31, Dr. Lillihei developed a rare parotid gland tumor and was told he had only a 5–10% chance of 5 year survival [23]. As most of you know, Dr. Walt Lillihei went on to become a founder of cardiothoracic surgery and heart-lung bypass and lived to be 80 years old. Although we must use available population level data to inform clinical decisions and care plans, we do not know “which patient” we are addressing. Often we, both patients and clinicians, seek certainty and clarity, all too easily forgetting humility in front of life. We cannot bear the thought that we are out of control. Yet, isn’t it arrogant to think that we know what’s going to happen even today, not to mention for the next 25 years of someone’s life? Thank goodness, we are not in control, given that our perspective is so limited. Leaving open possibilities allows patients to create meaning throughout their lives, which often matters more than any medical “truth” which becomes manifest over time.

It has been 39 years since the creation of the Norwood procedure (1981), 52 years since the Fontan (1968), 62 years since the Glenn (1958), and 76 years since the BTT Shunt (1944). Over those decades,



Fig. 7. Alicia and Kenny Wilmoth with their beautiful children (from left to right): Nola (age 6), Landen (age 2), and Liam (age 6).



Fig. 8. Taylor Houlihan addressing her graduating high school class as valedictorian.

staged surgical palliation and improvements in lifelong medical management have positively affected the trajectory and quality of life for those living with single ventricle congenital heart differences. We continue to build upon the foundational work of those who lived decades before us to improve life for generations of patients to come. Today's unknowns afford us tremendous opportunities for impact in research areas unimaginable to our predecessors, such as tissue regeneration. As clinicians, we have the opportunity right now to offer individuals with Fontan circulation and their families the gift of "Yes": yes to a full life, yes to a pursuit of passions, yes to future accomplishments, and yes to an empowered existence. Instead of narrowly focusing on the failures and difficulties of single ventricle circulation which may color our perspective especially when working only with inpatients, we may be of more service if we reflect on the triumphs and full breadth of the lives of these exceptional individuals.

"Yes" starts with each of us. Let's flip the script, using small shifts in language to match our deepest intentions of compassionate service. Try the power of the positive today- we believe it will uplift the entire community.

Declaration of competing interest

None to disclose.

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