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RESEARCH ARTICLE

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Barriers to and facilitators of physical activity in pediatric pulmonary hypertension

Catherine M. Avitabile[1,2,3] | **Caroline O'Brien**[4] | **Divya Dureja**[1] | **Dana Albizem**[1] | **Jena Mota**[1] | **Melissa Xanthopoulos**[5,6,7] | **Babette S. Zemel**[2,8] | **Peter F. Cronholm**[4,9,10]

[1]Division of Cardiology, Children's Hospital of Philadelphia, Philadelphia, PA

[2]Department of Pediatrics, University of Pennsylvania Perelman School of Medicine, Philadelphia, PA

[3]Center for Pediatric Clinical Effectiveness, Children's Hospital of Philadelphia, Philadelphia, PA

[4]Mixed Methods Research Laboratory, University of Pennsylvania Perelman School of Medicine, Philadelphia, PA

[5]Division of Pulmonary and Sleep Medicine, Children's Hospital of Philadelphia, Philadelphia, PA

[6]Department of Child and Adolescent Psychiatry and Behavioral Sciences, Children's Hospital of Philadelphia, Philadelphia, PA

[7]Department of Psychiatry, University of Pennsylvania Perelman School of Medicine, Philadelphia, PA

[8]Division of Gastroenterology, Hepatology, and Nutrition, Children's Hospital of Philadelphia, Philadelphia, PA

[9]Department of Family Medicine, University of Pennsylvania Perelman School of Medicine, Philadelphia, PA

[10]Center for Public Health, Leonard Davis Institute of Health Economics, University of Pennsylvania, Philadelphia, Pennsylvania

Correspondence
Catherine M. Avitabile MD, Division of Cardiology, Children's Hospital of Philadelphia, 3401 Civic Center Blvd, 8NW49, Philadelphia, PA, 19104, USA.
Email: avitabilec@chop.edu

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Abstract

Therapeutic exercise has not been widely adopted in pediatric pulmonary hypertension (PH), despite adult data supporting its safety and efficacy. While physical limitations may prevent children with PH from participating in physical activity, other barriers to and facilitators of physical activity are unknown. Youth ages 8-18 years with World Symposium of PH diagnostic Groups 1-4, functional class I or II, and ambulatory status were prospectively enrolled in a cross-sectional study including separate 30-min participant and caregiver interviews regarding attitudes toward physical activity and a proposed exercise intervention in pediatric PH. Interview questions were guided by Social Cognitive Theory and explored autonomy, self-confidence, and self-efficacy. Interviews were transcribed, coded, and analyzed using an iterative process to determine themes and patterns. Demographics and relevant PH condition-specific data were abstracted from the medical record. Thirty PH participant/caregiver dyads were interviewed. Facilitators of physical activity included enjoyment/interest in the activity, socialization, incentivization, and feelings of safety and normalcy. Barriers to physical activity included lack of interest, fear/anxiety, and self-consciousness. Findings were

similar in children and adults. Participants rarely reported restriction of activity by caregivers or medical providers. Attitudes toward engagement in a proposed exercise program were generally positive and reflected personal experiences with physical activity. Monitored exercise interventions that focus on patients' interests, cultivate confidence, respect limitations, and acknowledge the need for extrinsic incentivization may have benefits in pediatric PH. Future trials should test the impact of these characteristics on patient well-being and clinical outcomes.

KEYWORDS

exercise, interviews, pediatric pulmonary hypertension, physical activity, qualitative research

INTRODUCTION

Pediatric pulmonary hypertension (PH) is a life-threatening heart and lung condition in which abnormalities of the pulmonary arteries lead to right ventricular cardiac dysfunction, heart failure, and high risk of death. Most patients report exercise intolerance which results in lower self-reported quality of life scores.[1,2] Children with PH engage in less physical activity than peers,[3] which is concerning since supervised exercise training safely improves exercise performance, hemodynamics, and quality of life in adults with PH.[4–13] Professional societies now recommend supervised, monitored exercise training as an adjunct to medications in adults with PH.[14] Pediatric exercise trials are limited. One small study demonstrated improved exercise performance after a 16-week home exercise program including cycle ergometry and resistance band exercises.[15] With these limited data, the characteristics of the optimal pediatric PH exercise program are unknown. Duration, type, and setting of exercise rehabilitation have varied among adult trials. Patient adherence has been inconsistently assessed. In youth, practical concerns of developmental differences, access to facilities or equipment, and parental time commitment must be considered as home programs may be critical to avoiding missed school and workdays. Finally, existing exercise interventions in youth with PH are not grounded in a specified behavioral theory to encourage behavioral change.[15] Social Cognitive Theory (SCT) is a behavioral theory that focuses on fostering self-efficacy, emphasizing goal setting, and accomplishing gradual changes.[16,17] SCT has been applied to physical activity interventions in healthy children and those with chronic diseases, including congenital heart disease.[18–22] As youth with PH face similar functional challenges to those with congenital heart disease, SCT may be a powerful framework in which to encourage self-confidence around physical activity.

As understanding patient and family perspectives is critical to informing future exercise interventions in pediatric PH, the aim of this study was to use semi-structured qualitative interviews to explore attitudes toward physical activity, investigate key behavioral constructs of SCT, and study the feasibility, acceptability, sustainability, and fidelity of an exercise intervention in pediatric PH.

METHODS

Youth ages 8-18 years with World Symposium of PH diagnostic Groups 1-4, functional class I or II, and ambulatory status were prospectively enrolled in a cross-sectional study. At least one parent or caregiver was required to be interviewed as well. Patients with single ventricle physiology were excluded.

Study team members with expertise in SCT (M.X., P.F.C.) developed the semi-structured interview guide (Supplemental files 1 and 2) based on a review of the literature, and clinical and methodological expertise. Questions explored autonomy, self-confidence, and self-efficacy, central to SCT.[16,17] Guided by Proctor's framework for elements that are key to the uptake and sustainment of an intervention,[23] questions also explored the feasibility, acceptability, sustainability, and fidelity of a proposed exercise intervention in pediatric PH. Separate 30-min participant and parent/caregiver audio recorded interviews were performed using the interview guide and transcribed verbatim. Non-English language speakers were interviewed with an interpreter, and the interpreted English responses were transcribed. Two qualitative researchers analyzed the transcribed interviews using an integrated approach to deductively assess a priori SCT constructs after open coding of transcripts.[24]

The two coders double coded six transcripts (29%) and resolved discrepancies in their coding until consensus was reached ($\alpha > 0.75$). After the inter-rater reliability was completed, the remaining transcripts were independently coded by one coder (C.O.).

Demographics, medications, and relevant PH condition-specific data were abstracted from the clinical record. Standard descriptive statistics [mean ± standard deviation or median (interquartile range)] were used to summarize clinical characteristics.

RESULTS

Thirty PH participants and their parent/caregiver met inclusion criteria, and 60 interviews of participant and caregiver dyads were completed, reaching thematic saturation. Demographic and clinical characteristics are shown in Table 1.

Facilitators of physical activity

Enjoyment

A common facilitator of physical activity was enjoyment of a certain activity. There was variation in the type and intensity of activity that participants liked, but there was consensus that youth were motivated to do the physical activities they enjoyed. When asked about their ideal exercise program, participants typically referenced activities they had reported appreciating earlier in the interview.

> *"[My ideal program] would definitely have rock climbing."*
>
> (Patient Participant 3)

> *"Ideal exercise program. I mean, I would say biking, because he likes riding his bike, but not necessarily stationary bike. I couldn't even fathom him on a stationary bike."*
>
> (Caregiver Participant 7)

> *"Easier for me to exercise? I don't know. Probably when -- I mean if I find it interesting, I'll do it on my own time."*
>
> (Patient Participant 10)

Caregivers also noticed that children tended to prefer activities that did not call attention to their physical

TABLE 1 Child participant demographic and clinical data (N = 30).

Variable	n (%) or median (IQR)
Age at interview, *years*	14 (11, 16)[a]
Female	17 (57)
Race	
White	20 (67)
Black/African American	9 (30)
Asian	1 (3)
Hispanic or Latino	2 (7)
WSPH classification	
Group 1 – Pulmonary arterial hypertension	17 (57)
Group 2 – PH due to left heart disease	2 (7)
Group 3 – PH due to lung disease	10 (33)
Group 4 – Chronic thromboembolic PH	1 (3)
WHO Functional Class	
I	15 (50)
II	15 (50)
Number of medications	2 (1, 3)
BNP, *pg/mL*	28.2 (10.9, 50.7)
6MWD, *meters*	591 (495, 640)
Echocardiographic data	
Interval from study visit, *days*	0 (0, 70)
TAPSE Z-score	−2.3 (−5.1, 0.1)
RV fractional area change, *%*	37.9 (30, 42.2)
RV global longitudinal strain, *%*	22 (19.1, 23.6)
RV free wall strain, *%*	24.5 (21.4, 28.6)
RV/LV ratio	0.8 (0.7, 1.0)
LV eccentricity index	1.1 (1.0, 1.2)
Cardiac catheterization data	
Interval from study visit, *months*	8 (17, 44)
Mean pulmonary artery pressure, *mm Hg*	27 (20, 34)
Cardiac index, $L/min/m^2$	3.9 (3.5, 4.1)
PVRi, $Wood\ unit*m^2$	4.4 (2.8, 6.6)

Abbreviations: BNP, brain type natriuretic peptide; IQR, interquartile range; LV, left ventricle; PVRi, indexed pulmonary vascular resistance; RV, right ventricle; TAPSE, tricuspid annular plane systolic excursion; WHO, World Health Organization; WSPH, World Symposium of Pulmonary Hypertension; 6MWD, 6-min walk distance.

[a]Age range of enrolled participants was 8-18 years.

limitations, instead preferring to continue with activities they already felt confident doing.

> *"... [Child] always lags behind physically. So other kids when they're outside doing Tee-ball or playing soccer at four, five, and 6 years old, [Child] didn't do that. So, he's just behind physically. And then now I think he has realized at this age that he's not like those other kids. So, there's some things that he chooses not to try. And then we always force him to do things to look like other kids because I don't think he's going to look like other kids physically if that makes sense."*
>
> (Caregiver Participant 8)

Caregivers echoed the sentiment that at least some activities in an ideal exercise program would be familiar to their child, but it should also include new activities to build self-confidence.

> *"But if we're talking about [an ideal program] particularly for [my child] as well would be like get out of our comfort zone. We're going to try like three pounds. We're going to do something like strength training or we're going to do something like yoga or we're going to do something like boxing. I don't know. So, I would say like predictable and consistent with, like that added like newness or spontaneity."*
>
> (Caregiver Participant 13)

Socialization

Not every participant liked to exercise in groups, but participants commonly reported using physical activity as a social outlet. Participants mentioned they were willing to engage in more intense physical activity or simply enjoy it more when they were able to exercise with friends.

> *"Social part [of exercising] is huge and I also think now that she's just getting more involved in school, most of her good friends are there rather than at dance. I really don't know if she's going to go back to dance, she's happy enough doing the play or managing one of the teams or whatever, just be with her friends."*
>
> (Caregiver Participant 2)

> *"I guess having a friend, just not exercising by yourself, [it gets] more boring..."*
>
> (Patient Participant 5)

> *"I think when he's with a group of his peers... or like in gym class he'll participate or all the guys are going to go to Urban Air, he'll go... Yeah, when there's a little healthy peer pressure."*
>
> (Caregiver Participant 5)

Motivation/encouragement

Some caregivers reported positive feelings about themselves as parents when watching their child safely engaging in physical activity. They reported satisfaction seeing their child meet or push their physical limits without hurting themselves.

> *"I think the first thing that comes to mind [when thinking about them exercising] is... sorry I'll probably cry a lot. I feel very grateful. Because I didn't think that she would be able to have the level of exercise that she currently does. And if we're going to go back but initially with some apprehension and fear like, is this too much? Is something going to happen. But now just a tremendous amount of just gratitude I think or help or joy pride."*
>
> (Caregiver Participant 13)

> *"I do feel good when I see him doing the best of his ability. Not that he has to necessarily keep up with other kids, but when I see that he's trying his best, it makes me feel good, because I'm not putting him in that bubble. [...] And to see him out there [...] exerting himself to as much as his abilities, it makes me feel good as a mom, although of course the fear sets in of everything else, but that he's trying his best and he is pushing to his limits, and we're not sheltering him."*
>
> (Caregiver Participant 7)

Self-motivation

Participants were not generally motivated to be physically active for the sake of their health and wellness,

though the older participants reported that seeing improvements in their abilities would motivate them to continue with an exercise program.

> *"Seeing that she can walk further than she did, that she can tell she's progressing rather than staying the same or maybe getting worse. Anything like that really motivates her."*
> (Caregiver Participant 9)

> *"... Feeling better in everyday life. Just maybe building up a little more stamina. Just say progressively getting better at just running and building up that. Just maybe being a little bit stronger, nothing crazy. I don't expect to be become some athlete or something but just feeling better in general because I do feel I exercise more I probably would feel better."*
> (Patient Participant 9)

However, participants more commonly reported being motivated by external rewards and opportunities to socialize and play.

> *"What motivates me to do my homework is when my parents let me play video games."*
> (Patient Participant 3)

> *"Really at this point what motivates him is money or computer time."*
> (Caregiver Participant 5)

Feelings of normalcy/safety

Several caregivers commented that their child did best in situations where they were not singled out for having a medical condition, but still received support and medical attention when necessary. Multiple caregivers emphasized the importance of other adults allowing their child to stop exercising when they said they needed to stop.

> *"I think she would like to know that there's other kids that also have limitations definitely because I think sometimes the trouble she's running to is just not wanting to be different and so you don't say out loud. [...] She would need to know they know if you leave the room or if you stop or whatever, they're fine with it."*
> (Caregiver Participant 2)

> *"So, us being aware of that and being able to relay that to gym teachers has always been helpful, just that they're aware to let him listen to his own body."*
> (Caregiver Participant 5)

Several participants commented that activity is easier when they are feeling generally healthy and rested, beyond their PH symptoms being managed.

> *"If I'm like super hydrated that day, it's a lot easier. Also, like for dance, it got easier over time because I was just used to doing that stuff, but now if I were to go back to dance, it would probably be harder. So, if I'm used to doing something or I know how to do it, then it's a lot easier."*
> (Patient Participant 2)

Barriers to physical activity

Restrictions

Participants rarely reported restriction of activity by caregivers or medical providers. One participant mentioned their parents might restrict them more if they were more active, but they did not have enough interest in exercise for that to be relevant. More frequently, participants mentioned barriers such as feelings of self-consciousness, fear, anxiety, or a lack of interest.

> *"[My parents] don't want me doing stuff that's like super active. If I were to say like I want to go for a run, they'd probably say, 'Don't do that.' But yeah, like, I don't want to go for a run."*
> (Patient Participant 2)

Fear/anxiety

Caregiver feelings were heavily dependent on the severity of the patient's PH or the occurrence of a recent medical event. Caregivers of high functioning/asymptomatic

patients reported fewer feelings of anxiety about their child exercising.

> *"I think considering where we are, he is not in any danger of overdoing it. So no, I don't worry."*
>
> (Caregiver Participant 5)

Comparatively, caregivers of children with more severe PH expressed more concern and fear around their child exercising.

> *"Nervous. I think that exercising is a good sign, that [Child]'s feeling good enough to exercise. But at the same time it makes me a little bit nervous that [Child]'s heart might get strained with exercise or vigorous activity. So it's kind of a mixed bag... Running makes me nervous. Any activity that I know [Child]'s heart rate's going to be high makes me nervous."*
>
> (Caregiver Participant 15)

Participants had a similar relationship between the severity of their symptoms and anxiety with physical activity. One patient reported feeling anxious when getting short of breath.

> *"Like the other day, I was chasing somebody. I got super out of breath, and like I felt super scared because like it did feel like something was gonna happen..."*
>
> (Patient Participant 2)

Comparatively, an asymptomatic participant reported only feeling stressed while exercising when they were getting ready for a competition.

> *"[I get nervous] Before a meet, because I'm trying to win."*
>
> (Patient Participant 1)

Lack of interest

Like adults, children with PH are more motivated to participate in physical activity that they enjoy and feel confident doing. There were some participants who reported not being very physically active, but also added that sports and exercise simply were not interesting to them.

> *"I don't really play any sports, but in gym class when we have to, I play volleyball, basketball. I'm very short."*
>
> (Patient Participant 3)

> *"He's in high school, so they don't have recess. Last semester he had PE, but this semester he does not. So yeah, like whatever walking around the classroom and back and forth to classes, that's pretty much it. He refuses to walk the dog or clean the room. He just wants to be on video games and computers all the time."*
>
> (Caregiver Participant 5)

Self-consciousness

The most frequently cited barrier to regular physical activity was self-consciousness. Both participants and their caregivers commented that a child's physical limitations made them stand out from their peers, leading to negative feelings when exercising.

> *"[It's harder to exercise] If somebody is watching me, maybe it makes me a little uncomfortable. I don't know, I feel like it's just anything though. Like if somebody is watching me run around."*
>
> (Patient Participant 12)

> *"Honestly, when it's more like with just him not necessarily doing like a team, like it's - easier?... It's like when he is out there, he does limit himself like when he does get out of breath, so he'll stop or take himself out or sit down or whatever it is. He's kind of gotten used to just doing things by himself or with one other person that doesn't mind matching his level of exercise."*
>
> (Caregiver Participant 8)

> *"And she's not as fast as the rest of the kids. So, but that kind of slows her down. Like, I know, outside playing tag -- she's always it because she, she is the first one that they catch. So that, that kind of discourage her, she, like playing tag and because she doesn't*

move as fast, she sometimes wants to sit out."

(Caregiver Participant 22)

Standing out from peers was a significant worry about exercising. No participants said they were bullied or socially ostracized because of their PH, but both participants and caregivers reported feeling different from peers as a deterrent from regular physical activity.

"I know that in tennis that the other kids you know continued to play harder and longer and whatever, but he was getting out of breath. My husband is saying to me he was getting out of breath you know that probably had something to do with [him quitting]."

(Caregiver Participant 10)

"I actually avoid [exercising] in the first place and I don't exercise if other people are around, so I don't go to the gym and exercise because I'm self-conscious about getting more out of breath and people and just little things like that."

(Patient Participant 15)

"Like if I was playing basketball with people that are actually good at basketball, I wouldn't feel confident even though like I'm okay at basketball and I wouldn't feel confident doing it, but if I'm doing it with people that are more my level, then I would feel a lot more confident."

(Patient Participant 6)

At least one participant explicitly said struggling with physical activity deterred them from participating in it and that they enjoyed exercising less when their PH symptoms were activated.

"Just in the past I know that when I run I can't really do it for that long and I get really out of breath and tired. So I don't really enjoy doing it and that's why I don't really feel confident when I do it because I normally just (get) really out of breath quickly."

(Patient Participant 17)

Reactions to a hypothetical exercise program

Patients and their caregivers were asked about the patient participating in a regular exercise program (defined in the interview guide as "regular days and times"). Participant responses were generally positive but varied in their reactions to specific components of a program. Specifically, attitudes toward scheduling, activity selection, and motivation varied.

Scheduling

Scheduled exercise was both a barrier and a facilitator of program participation, depending on the temperament of the patient participant. Some participants preferred to choose the timing of their exercise and communicated that scheduled exercise would discourage them from giving full effort. Other participants reported they were already engaged in many scheduled/organized activities, and they liked the routine. More active patients said they would not be interested in adding another activity to their already full schedules.

"I kind of wouldn't want to, because I like to exercise and stuff when I'm feeling it. Like, if I'm not feeling it, I don't really do it and I don't go my full amount, like, I don't do the best I could do if I'm not feeling it."

(Patient Participant 26)

"I mean, since my usual schedule is Monday I have karate, and then Tuesday I play with my friends, and then Wednesday I have my second karate, and then Thursday is baseball practice, and then Friday, I'm just relaxing, so I'm kinda used to that. So I think I'd be fine [not doing more]."

(Patient Participant 30)

Ideal activities

Almost universally, participants said the best activities for the hypothetical exercise program were activities they or their child already enjoyed. Participants who enjoyed walking wanted a walking program. Participants already in a baseball league wanted to play baseball. The patient who liked rock climbing wanted to

keep rock climbing. Conversely, participants wanted the option to decline activities they were afraid of or did not enjoy.

> *"So, she still can't ride a two wheel, bike. We tried but there's the fear factor for her as well with that. I'm not sure if that gets physical but she is, her balance isn't great for that sort of thing that her reaction times I think, something she does want to do, she does want to do certain exercises. So, bike riding would be out. What would be a good exercise for her though? I can't think of anything else. I've been walking, walking the dog. Go out to shops, rock climbing and we have a membership."*
>
> (Caregiver Participant 3)

Motivation

Like their current physical activity routines, patients and caregivers cited a variety of external and internal motivators for continued participation in a hypothetical exercise program. Some participants reported enjoyment would be sufficient motivation, while others would be willing to increase their physical activity in return for more access to video games/screen time or a higher allowance. Still others said they would want to see growth and increased performance to feel motivated to continue with the program.

> *"I'm not that good at riding my bike uphill, so maybe I -- and I really wanna know how to ride uphill better, so maybe that motivates me."*
>
> (Patient Participant 31)

> *"At this stage, money. Obviously it's part of the reason he likes the research studies because he gets the ClinCards."*
>
> (Caregiver Participant 8)

When discussing willingness to engage with the hypothetical program, feelings of normalcy were frequently cited again. Caregivers emphasized that any program that made their child feel singled out or program leaders that did not respect the limits of a patient's abilities would be very discouraging to patients.

> *"I guess, just whoever is leading it isn't too pushy. And again, I don't think that would be the case for something like this, but, as long as they're patient and understanding, again, I think, they would have to be for something like this, though. I think that would make him feel comfortable. Sometimes, he's pretty unsure of himself for certain things, especially if it's new, like a certain activity or whatever. So, he can be pretty hesitant.*
>
> (Caregiver Participant 23)

> *"I think she would like to know that there's other kids that also have limitations definitely because I think sometimes the trouble she's running to is just not wanting to be different and so you don't say out loud [...] I think having other kids that would understand that would be really big. She definitely needs to know that the adults surrounding anything fully understand. She's just a very compliant kid. She would need to know they know if you leave the room or if you stop or whatever, they're fine with it."*
>
> (Caregiver Participant 2)

DISCUSSION

Despite adult data supporting the safety and efficacy of exercise rehabilitation in PH, therapeutic exercise has not been widely adopted in pediatric patients. Barriers may include lack of clinical trial data, fear of adverse events, limited resources, physician restriction, or inadequate study of the characteristics of a successful physical activity program in this population. This is the first pediatric study to collect perspectives on physical activity and exercise programming from patients and families, which are critical to designing and implementing successful therapeutic interventions. For children, facilitators of activity included enjoyment, socialization, motivation, and conditions that promoted safety and normalcy. On the contrary, children with PH were limited by disinterest, fear or anxiety, and self-consciousness. Integrating these rich qualitative data on barriers to and facilitators of activity in pediatric PH are critical to informing future exercise interventions in this population.

Several adult PH studies have reported patient perspectives on exercise. Chia studied 187 PH patients from 19 countries and identified barriers including: fear, frustration, and uncertainty; symptoms of dyspnea and fatigue; and cost and access to appropriate services.[25]

When asked about the characteristics of an ideal multidisciplinary rehabilitation service, participants stressed education, supervision, structure, and psychology input as crucial components. In a smaller, single-center United States study, Cascino reported barriers to activity including lack of interest, lack of energy, and lack of self-discipline.[26] The authors suggested that physical activity interventions should address interest, enjoyment, and skill development. McCormack studied attitudes toward exercise in 19 adult patients in Ireland and identified three themes including fear, environmental factors, and perceived (lack of) value of exercise, suggesting that future interventions that mitigate fear and educate patients on the value of exercise may have positive impact on engagement.[27] Finally, in a separate study, McCormack investigated PH patients' knowledge of physical activity, exercise preferences, and support needs.[28] The investigators found that patients felt they received suboptimal activity advice from their clinicians, yet they desired that guidance. Patients also preferred to exercise in a monitored home setting with a system to support external accountability and facilitate continued engagement.

In our study, children with PH and their caregivers expressed some similar attitudes as adults, but they also displayed some notable differences. Like adults, children are motivated to engage in physical activity when they can choose an activity they like. Parents of children with PH suggested a combination of familiar and new activities may give children more options and increase engagement. Families stressed that programs must address needs and abilities and identify activities that work for an individual child. Semi-structured, developmentally appropriate programs may be needed to sustain engagement in children with PH across age ranges. Children were generally not motivated to exercise based on the perceived value of physical activity but reported needing external motivation and incentivization. Goal setting with rewards for achievement may be a central component to pediatric programs. Some participants were motivated by opportunities for socialization within a group setting, but this would likely be most powerful if the group only included other PH patients or activities that did not socially isolate children with activity limitations.

Children and families with PH want to feel safe and appropriately monitored in an exercise program. There were some reports of fear of over-exertion. However, there was some disconnect between provider clinical assessment and reported symptoms. Study participants were all functional class I or II – asymptomatic or minimally symptomatic from their PH. Participants rarely reported activity restriction from their healthcare providers. Yet, participants and caregivers with "more severe PH" expressed more concern and fear around their child exercising. The roots of these fears are not clear and future studies should explore attitudes of pediatric PH providers toward exercise and how these impact recommendations. Even beyond the need for safety, PH patients expressed a need to feel "normal" when exercising. The most common barrier to regular physical activity was self-consciousness. Physical limitations made children with PH stand out from their peers, leading to negative feelings when exercising. Patients and families focused on the need to build self-confidence and alleviate self-consciousness. When limitations were evident, participants stressed the need to stop exercising. However, this did not always seem to be due to perceived danger, but rather a desire to return to a state of control.

Multiple other recent interventions have used SCT to encourage physical activity in healthy youth and those with chronic health conditions.[18–22] This approach has not been applied in pediatric PH, but fostering self-efficacy is one of the most powerful factors in predicting behavior in line with SCT.[16,17] Digital health nudging via smart phone messages and individually tailored e-Health encouragements based on SCT have been studied to increase physical activity, self-efficacy, and health related quality of life in teenagers with congenital heart disease, a population with similarities to pediatric PH.[18,19] When Willinger used smart phone messaging to encourage physical activity measured by wearable tracker in patients with congenital heart disease, emotional wellbeing increased.[18] Stephens demonstrated the feasibility of a 12-week mobile application physical activity intervention informed by SCT including coaching, monitoring of physical activity by wearable device, goal setting, social support, and education in youth with multiple sclerosis.[20] The intervention was feasible, and adherence was high. While physical activity did not increase, SCT scales demonstrated increased social supports and decreased outcome expectancy. These and other studies demonstrate that more work is needed to understand how to incorporate aspects of SCT approach into physical activity interventions in youth. When we asked, youth with PH and their caregivers questions focused on self-efficacy and self-confidence, they responded that they were motivated to participate in activities when they felt confident and the most common barrier to physical activity was self-consciousness. This may suggest that physical activity interventions with interval goal setting to foster self-efficacy, increase competence, and instill confidence could increase intrinsic motivation to adhere to a program. Findings from this study have informed an on-going home physical activity intervention in pediatric PH.[29] Elements of the intervention that are based in SCT and the themes expressed in these interviews include: 1)

semi-structured exercise prescriptions tailored to the participant's baseline level of activity, interests, and access to resources; (2) interval goal setting fostering self-efficacy; (3) a digital platform enabling continued communication and feedback between participant and study team; and (4) multiple avenues to assess participant safety.

There are some limitations to this study. Only patients in functional classes I and II were enrolled as these findings were meant to inform a pilot, home physical activity intervention in those patient groups. More symptomatic patients and their caregivers may have different attitudes toward physical activity, but they could also benefit from a supervised exercise program as part of a comprehensive treatment plan. However, it is possible that these themes – need for enjoyment, safety, normalcy, minimal self-consciousness – may resonate with more symptomatic patients as well, and medically-appropriate exercise guidance could incorporate these perspectives across the pediatric PH spectrum. This study was designed to obtain preliminary data across a range of ages and genders. We did not have enough sample size to explore to saturation differences in attitudes by subgroups of patients (i.e. children vs. teens; male vs. female vs. other genders; active vs. sedentary patients). Future, larger studies should explore any demographic, social, or environmental factors that impact attitudes towards activity and the uptake of a proposed exercise intervention. Participation bias resulting from patients who declined the study may have provided valuable additional perspective. Social desirability may have shaped some responses. The semi-structured interview guide was specifically created to investigate autonomy, self-confidence, and self-efficacy and to explore characteristics of a proposed exercise intervention in pediatric PH. Responses focused on these attitudes and elements. However, it is unknown whether SCT is a comprehensive framework on which to base an exercise intervention in pediatric PH. This should be explored in future pilot interventional studies. Finally, this study did not include provider recommendations or attitudes toward physical activity or exercise prescription. While a minority of participants reported provider exercise restriction, attitudes and influence of the medical team are not addressed here.

In summary, youth with PH and their caregivers endorsed facilitators of physical activity and organized exercise including enjoyment, motivation, and feelings of normalcy. Disinterest, fear or anxiety, and self-consciousness impeded participation in physical activity and could impact the successful implementation of an exercise program. Monitored interventions that focus on patients' interests, cultivate confidence, respect limitations, and acknowledge the need for extrinsic incentivization may have benefits in pediatric PH. Future trials should test the impact of these characteristics on patient wellbeing and clinical outcomes.

AUTHOR CONTRIBUTIONS

CMA designed the study and crafted the manuscript. CO performed all interviews, coding, and analyses and edited the manuscript. DD, DA, JM, and AB provided collaboration in study design and execution and edited the manuscript. MX, BSZ, and PFC provided senior mentorship in study design and execution and edited the manuscript.

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CONFLICT OF INTEREST STATEMENT

The Authors declare that there is no conflict of interest.

ETHICAL STATEMENT

Fully informed, written consent was obtained from the parent/legal guardian of participants <18 years and of participants 18 years of age. In addition, age-appropriate informed assent was obtained from participants <18 years. This study was approved by the Children's Hospital of Philadelphia Institutional Review Board protocol #20-017976.

GUARANTOR

Catherine M. Avitabile MD.

ORCID

Catherine M. Avitabile http://orcid.org/0000-0002-2354-5447

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SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

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