Pediatric Cardiac Rehabilitation: a Review

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Abstract

Purpose of Review In addition to concerns with physical health and activity levels, children with cardiac conditions can be at risk of neurodevelopmental and socioemotional maladjustment. Children with congenital heart defects requiring surgery early in life are at risk of developmental delays and cognitive impairments, and both children with congenital heart defects and those with cardiomyopathies are at risk of socioemotional concerns. As a result, there is an increasing focus on rehabilitation efforts for these patients, in order to improve both their physical well-being and their adjustment outcomes. However, there are no established standards for rehabilitation programs applicable across children with cardiac conditions, in stark contrast to guidelines for adult patients. The purpose of the present review is to summarize recent studies on pediatric cardiac rehabilitation and describe the structure of our own program, in order to aid with the delineation of future guidelines.

Recent Findings Twenty programs for pediatric cardiac rehabilitation were identified and reviewed. We review inpatient, outpatient, and home-based programs, most of which include two to three sessions of exercise training per week for 12 weeks with a focus on improving exercise capacity. We also review emerging cognitive rehabilitation for children with cardiac disorders and discuss a newly developed program at our own institution.

Summary A review of past findings, along with recent efforts at our institution, suggests that a structured cardiac rehabilitation program can benefit children by increasing exercise response and physical activity as well as improving developmental, cognitive, and psychosocial outcomes.

Keywords Pediatric cardiac rehabilitation · Congenital heart defects · Cardiomyopathies · Heart transplantation · Neurodevelopmental outcomes · Socioemotional adjustment

Children with cardiac disorders are at risk of neurodevelopmental and socioemotional maladjustment [1–4], on top of concerns with their physical health and activity levels. As a result, there is an increasing focus on rehabilitation efforts for these patients, in order to improve both their physical well-being and their psychosocial adjustment. However, there are no established

standards for rehabilitation programs applicable across children with cardiac conditions, in stark contrast to guidelines for adults [5]. In this review, we summarize recent studies on pediatric cardiac rehabilitation and present the structure of our own center's program, in order to aid with the delineation of future guidelines for pediatric patients.

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Pediatric Cardiac Disorders

Congenital Heart Defects

Congenital heart defects (CHDs) are one of the most common birth defects, affecting approximately 1.35 million newborns worldwide every year [6] and 1 million children of all ages in the USA [7]. CHDs encompass a wide range of conditions in which there is abnormal development of the heart and/or its major blood vessels; they are broadly categorized into cyanotic defects (where oxygen-rich and oxygen-poor blood mix)

and acyanotic defects (where the amount of oxygen delivered throughout the body is unaffected). A summary of heart defects is provided in Table 1, with abbreviated details on clinical presentation and management.

Children with CHDs can have comorbid pulmonary and other medical complications hindering their ability to function [8, 9], and they are at risk of delays in motor development and exercise intolerance [10, 11]. Impairments in motor functioning and exercise tolerance, in turn, have been linked with perioperative morbidity and sedentary behaviors (which can have negative health consequences) [12]. Along with their physical impairments, children with CHDs are more likely to have parents, educators, and health providers who excessively limit their activity, predisposing them to physical inactivity and exercise intolerance [13]. They may, therefore, benefit from cardiac rehabilitation with an

emphasis on increasing physical activity and providing relevant education.

Children with CHDs are also at increased risk of early developmental delays, later cognitive dysfunction, and poor quality of life, including speech and language delays, deficits in attention, executive functioning and visuospatial skills, and emotional and behavioral dysregulation [1, 2]. Neurodevelopmental disruptions are thought to result from a combination of brain dysmaturation [14–16] (which likely arises from genetic factors and teratogens that concurrently impact cardiac and cerebral fetal development [17, 18]), altered cerebral perfusion (which can then affect functional and structural brain development) [19], and neurological injuries (i.e., cerebral hemorrhages, ischemic lesions, periventricular leukomalacia, and other white matter injuries) [20, 21]. The complexity of the heart defect has been related to the severity of neurodevelopmental concerns, with children with

Table 1 Congenital heart defects and their clinical presentation and management

Congenital heart defect	% of cases with CHDs	Presentation	Management
Acyanotic Defects			
Atrial septal defects	7–10	Fatigue, but usually asymptomatic	Repaired surgically via patch or suture closure
Ventricular septal defects	50–60	Symptoms of overcirculation within the pulmonary vasculature, congestive heart failure (CHF), cardiomegaly on chest radiograph, and evidence of pulmonary vascular congestion	Management of CHF, management of nutrition, and ultimately surgical patch repair
Atrioventricular septal defects	5	Features of CHF, failure to thrive, cardiomegaly on chest radiograph, and evidence of pulmonary vascular congestion	Nutrition monitoring and management, diuretics, and ultimately surgical repair
Aortic and pulmonic valve stenosis	Aortic, 5–8 Pulmonic, 8–10	Systolic ejection murmur	Catheter-based balloon valvuloplasty, surgical valvuloplasty
Coarctation of the aorta and interrupted aortic arch	Coarctation of the aorta, 5–8 Interrupted aortic arch, 1.5	Lower post-ductal saturations, blood pressure gradients in upper and lower extremities, poor feeding, and cardiogenic shock if in crisis	Prostaglandin infusion to keep the ductus arteriosus open in newborns, surgical repair
Cyanotic Defects			
Tetralogy of Fallot	5	Hypercyanotic spells with agitation, and fever or illness	Surgical management, palliative procedures, complete repair including closure of ventricular septal defect and resection of right ventricle obstruction
Transposition of the great arteries	2	Cyanosis in the first 12 h of life, narrow mediastinal silhouette on radiograph	Prostaglandin infusion, surgical repair
Truncus arteriosus	2–5	Cyanosis symptoms of pulmonary overcirculation in the first few hours of life	Diuresis and fluid management, surgical repair
Total and partial anomalous pulmonary venous drainage	1	Respiratory distress and cyanosis, radiograph with white out of the lung fields and small heart, partial anomalous return possible in late childhood and adolescence	Surgical repair
Hypoplastic left heart syndrome	2–3	As the ductus arteriosus closes, infants show signs of shock with poor pulses and perfusion, poor urine output, pulmonary overcirculation, and radiograph with vascular congestion and cardiomegaly	Multi-stage surgical palliation including Norwood, Glenn, and Fontan procedures in infancy through 2–4 years of age

Summarized from Puri et al. [91]

single ventricle lesions thought to be at particular risk [1]. Even with simpler defects, though, factors such as cardiac arrest [22] and extracorporeal membrane oxygenation (ECMO) [23] can portend increased risk of neurodevelopmental delay. As such, children with CHDs may benefit from access to habilitation and rehabilitation services, during acute care following surgical interventions, cardiac arrest, and ECMO but also follow-up interventions for functional impairments or developmental delays that emerge throughout childhood.

Cardiomyopathies

Cardiomyopathies occur in fewer than 2 per 100,000 children [24, 25]. Cardiomyopathies are abnormalities of the ventricular myocardium that cannot be explained by abnormal loading conditions or heart defects; they can result from coronary artery abnormalities, tachyarrhythmias, exposure to infection or toxins, or other underlying disorders [26]. There are several subtypes of cardiomyopathies, summarized in Table 2 along with their presentation and management.

In many children with cardiomyopathies, the disorder progresses to the point where medications and surgical options are ineffective. Nearly 40% of children with symptomatic cardiomyopathy require a heart transplant or die [27, 28]. The time to transplant or death for children with cardiomyopathy has notably not improved over the past several decades, and the most economically advanced nations have no better survival outcomes than developing nations [29]. Rehabilitation programs may be beneficial, not to cure the diseases, but to potentially improve the cardiac function and the overall health of children for whom options for care are limited [30••].

Children with cardiomyopathies, particularly those who progress to heart failure, have an increased risk of anxiety, depression, and quality of life concerns [3, 31••]. In addition, those who require permanent ventricular assist devices or heart transplantation can have mild neurodevelopmental disruptions [4, 32]. Among adults, cardiac rehabilitation programs have yielded success in improving not only cardiac function and overall physical activity but also emotional well-being and quality of life. Children may similarly benefit from rehabilitation programs.

Heart Transplantation

Per the Registry of the International Society for Heart and Lung Transplantation [33], there are approximately 400 to 600 cases of pediatric heart transplantation per year, worldwide. CHDs are the most common indication for transplant during infancy (55% of cases), followed by cardiomyopathies (37%). However, for children over age 10, cardiomyopathies are the most common indication (43–54% dependent on age). The median post-transplant survival is approximately 22 years for those transplanted in infancy, approximately 18 years for those in early childhood (age 1–5 years), approximately 14 years for those in middle childhood (age 6–10 years), and 13 years for those in adolescence.

Cardiac transplantation results in postganglionic denervation, leading to the inability to respond to the parasympathetic nervous system [34]. In turn, there are higher systolic and diastolic blood pressures, elevated heart rate at rest, lower maximal myocardial oxygen consumption, lower heart rate reserve, and decreased exercise duration. Because of these physiologic changes, cardiac rehabilitation can be helpful in reestablishing physical health and activity levels.

Table 2 Cardiomyopathies and their clinical presentation and management

Cardiomyopathy	% of cases with cardiomyopathy	Presentation	Management	Survival
Dilated cardiomyopathy	> 50	Ranges from asymptomatic to acute decompensated heart failure and cardiogenic shock; arrhythmias	Medical therapies, mechanical support, and transplant	60–75% at 5 years for those without heart transplant
Hypertrophic cardiomyopathy	42	Ranges from asymptomatic with or without murmurs to exercise intolerance, chest pain, palpitations, syncope, or cardiac arrest; increased risk of sudden death with exercise	β -Blockade, calcium channel blockers, disopyramide, surgical myomectomy, and automatic implantable cardioverter-defibrillator	97% at 5 years
Restrictive cardiomyopathy	4.5	Ranges from asymptomatic to overt heart failure, syncope, or sudden death; arrhythmias	Diuretics, anticoagulation, antiarrhythmics, automatic implantable cardioverter-defibrillator, early consideration for transplant	68% at 5 years
Noncompaction cardiomyopathy	4.8	Benign or severe course with progressive systolic or diastolic dysfunction, life-threatening arrhythmias, or thromboembolism	Anticoagulation, consideration for transplant	Death or transplantation in 18–25%

Summarized from Lee et al. [26]

Heart transplantation in infancy and toddlerhood has been associated with mild delays in motor and cognitive development, with a similar pattern of deficits as seen with children with CHDs requiring surgery other than transplantation early in life [35]. Specifically, this group can display mild reductions in motor skills, intellectual abilities, language abilities, and visuospatial skills. Older children, who often undergo a heart transplant for cardiomyopathy, can display mild cognitive deficits and are, more importantly, at risk of internalizing and externalizing distress [36]. Among children who undergo heart transplantation, cardiac rehabilitation may be particularly important not only to help pediatric patients regain their strength and mobility in the short term but also to help maximize children's quality of life across their reduced lifespan.

Pediatric Cardiac Rehabilitation

Guidelines from the American Heart Association and the American Association of Cardiovascular and Pulmonary Rehabilitation recommended that cardiac rehabilitation programs for adults and older adults include several core components including a baseline assessment, management of health risk factors (e.g., diabetes, hypertension, lipid levels) and nutrition, exercise training and physical activity counseling, and psychosocial management [37–39]. Similarly, primary goals for pediatric cardiac rehabilitation are managing physical health and activity as well as socioemotional functioning. Given the key developmental tasks of childhood, such as gaining basic academic skills, there may be an additional focus on mitigating developmental and cognitive disruptions, which would not be a primary concern during adult cardiac rehabilitation.

Addressing Physical Health and Activity

Historically, there have been concerns regarding the adverse effects of physical activity in those with cardiac conditions. However, research has underscored that exercise is not necessarily contraindicated. For example, a large study among adults with CHDs demonstrated that the majority of sudden cardiac events in patients occurred at rest (69%), with 11% during sleep, and only 10% during exercise [40]. In fact, physical activity can be beneficial for those with cardiac conditions. There is overwhelming evidence that exercise training within adult cardiac rehabilitation promotes cardiorespiratory fitness, strength, flexibility, and metabolic health; reduces morbidity, mortality, and hospital admissions; and improves quality of life [41•, 42•]. Emerging research with pediatric populations with cardiac conditions similarly suggests that exercise training can have beneficial effects, although there is some variability in effects [43, 44].

Given its potential benefit, exercise training has been outlined as a key component of cardiac rehabilitation with adult patients [37–39], recommended in guidelines for certain pediatric cardiac populations (e.g., those with heart failure), and has consistently been included in emerging programs with children [30••, 43–45]. From prior reviews of cardiac rehabilitation programs for children with CHDs or cardiomyopathies [30••, 43, 44], it has generally been recommended that programs have a duration of at least 12 weeks, with two to three sessions per week, and sessions of at least 30 min (and up to 90 min). Programs should include aerobic, resistance, and flexibility training, with warm-up and cool-down periods. Training should be individualized based on the results of metabolic stress tests, cardiac biomarkers, echocardiograms, baseline resistance-training capacity, and past medical history. Notably, the intensity of aerobic exercise should be at a heart rate approximately equivalent to anaerobic threshold. The patient's progress should be reviewed at least weekly, and progressive increases should be made in the child's exercise workload as tolerated and when medically appropriate. Programs might also benefit from a 6-month maintenance period with two exercise visits per month, including a review of exercise logs. Both center-based and at-home training programs may be effective [30••].

More broadly, cardiac rehabilitation programs can provide education on appropriate physical activity in children with cardiac conditions. Physical activity has been promoted in guidelines from the American Heart Association and the Association for European Paediatric Cardiology for children with CHDs [46••, 47]. Specifically, guidelines for physical activity in children with CHDs underscore the need for at least 60 min of daily activity that is developmentally appropriate and enjoyable. Vigorous activity is recommended at least 3 days per week, and exercises for strengthening bone and muscle (e.g., high-impact and anaerobic burst exercises such as jumping) are recommended 3 days per week. However, children with certain heart defects (e.g., transposition of the great arteries, single ventricle conditions) may need to limit participation in the highest intensity activities, notably in competitive sports [48]. As for all children, screen time should be limited to no more than 2 h per day for children over the age of 5 years, and children under the age of 3 years should not have any screen time [46••, 47].

Guidelines from the American Heart Association and the European Society of Cardiology for individuals with cardiomyopathies also underscore the benefit of physical activity [49, 50, 51••]. Still, those with cardiomyopathies may have more restrictions in their activity. Intensive exercise programs and competitive sport may be contraindicated for certain individuals depending on their subtype of cardiomyopathy, history of symptoms, and findings on cardiac testing. More generally, patients should be advised to start exercise sessions with a warm-up period and end with a cool-down period; avoid burst

exertion, preferably avoid high-intensity free weight lifting due to the risk of injury with syncope; avoid exercising in adverse environmental conditions; and exercise only in environments equipped with an automatic defibrillator.

In addition to exercise training and counseling on appropriate physical activity, pediatric cardiac rehabilitation programs can help patients and their families establish a nutrition plan and better recognize and manage the symptoms of their cardiac condition. As such, programs can promote a healthy lifestyle and decrease the risk and the severity of future cardiovascular disease.

Addressing Cognitive and Socioemotional Functioning

Cardiac rehabilitation programs can help not only mitigate the physiological effects of cardiac conditions but also enhance the socioemotional and cognitive functioning of patients. By doing so, cardiac rehabilitation programs can help children, adolescents, and young adults transition back to their family and home life, peer groups, and school and work settings following hospitalization. Although such efforts are standard in programs with adult patients, recent reviews suggested that the same cannot be said of pediatric programs, even though the omission of mental health providers may decrease the success of cardiac rehabilitation [43, 44].

Psychologists, social workers, and other licensed mental health professionals can assess the psychosocial needs of pediatric patients and their families and monitor changes in needs throughout their rehabilitation [52]. For example, although patients have typically undergone a psychosocial evaluation prior to a heart transplant, their needs may change during the waiting period for a heart and subsequent recovery from surgery. Families may also be more likely to report sensitive information once they no longer need to be concerned that they may not be listed for a transplant because of their financial, social, or personal resources. They may also share important information once they feel more comfortable with their care team over time. As such, they may be more apt to discuss difficulty coping with their illness and medical care, negative mood, insufficient social support, and/or premorbid history of mental health concerns.

Having identified patients' needs, mental health providers can implement interventions to foster healthy behaviors and treatment compliance and address the distress associated with critical illness and lengthy hospital stays. For instance, motivational interviewing strategies can aid with building a desire to engage in adaptive behaviors, such as taking medications and following nutrition and exercise plans [53, 54]. Strategies from behavioral management therapy can target the behavioral manifestations of distress often seen in young children, and cognitive-behavioral and mindfulness-based therapies can effectively target anxious, irritable, and depressed moods among older children and

adolescent [55–57]. Behavioral and cognitive-behavioral therapies also offer models for chronic pain and poor sleep [58, 59]. The ability to consult psychiatric colleagues within cardiac rehabilitation programs further allows pediatric patients access to specialized psychotropic medication management when indicated.

Interestingly, psychological interventions might benefit physical health along with psychosocial health. In a meta-analysis of 23 randomized control trials involving adult patients with coronary heart disease, psychological interventions both reduced emotional distress and improved systolic blood pressure, heart rate, and cholesterol levels [60]. Similarly, a pediatric cardiac rehabilitation program with a stress management component improved physiological measurements, although the study did not separate out the effects of exercise training, health education, and stress management [61]. Research has also documented associations between physical and emotional health among youths who have completed cardiac rehabilitation [62] and between daily physical activity and mental health among children with CHDs [63].

Cardiac rehabilitation programs can furthermore offer an opportunity to identify and address disruptions in the acquisition of developmental milestones and cognitive abilities. Developmental assessments can identify delays in motor, language, and adaptive development, and neuropsychological evaluations can identify cognitive deficits [1]. Such testing can aid with treatment and transition planning (e.g., clarifying the need for speech and language therapy and outlining recommendations for an Individualized Education Program upon return to school).

Importantly, families of children with cardiac disorders have described the need for effective communication among health and mental health professionals, families, and schools as critical within rehabilitation programs [64]. By incorporating mental health providers, care coordinators, and school liaisons into comprehensive cardiac rehabilitation programs, the needs of pediatric patients can be better understood and addressed by the treatment team.

Rehabilitation Programs

As summarized in Table 3, we identified 20 cardiac rehabilitation programs described across 26 reports, with samples that included patients under age 18. Due to the limited number of studies and consistency in findings, we did not exclude programs that had both pediatric and adult patients. Similarly, we included both randomized control trials and investigations without control groups, due to the emerging nature of the literature. Programs described within case studies and unpublished manuscripts (e.g., theses, conference presentations) were excluded from the review. We only included reports written in English and papers published after 1990.

Table 3 Cardiac rehabilitation programs including pediatric patients

References	N	Patient population	Age (in years)	Program protocol	Program duration	Measures	Outcome/results
Inpatient Acute Care Rehabilitation							
Hollander [65]	14	Ventricular assist device	< 1–12	<ul style="list-style-type: none"> • 4–5 1-h sessions per week via a standardized care pathway • 8–10 therapeutic goals • Physical and occupational therapies 	N/A	Completion of pathway goals focused on age-appropriate development and activities of daily living	<ul style="list-style-type: none"> • 11/14 achieved all goals • No adverse events
McBride [66]	20	Hospitalized for heart failure (i.e., cardiomyopathy and failing single ventricle)	Mean 13 ± 3.2	<ul style="list-style-type: none"> • 3 1-h sessions • Resistance and aerobic exercises (treadmill or cycle ergometer) 	2–18 months	Exercise testing*	<ul style="list-style-type: none"> • Peak work rate and VO₂ significantly below percentile for age and sex
Outpatient Rehabilitation							
Balfour [61]	7	CHDs or acquired heart conditions	13–19	<ul style="list-style-type: none"> • 3 days per week in facility • 1–2 days per week home exercise • 30–40 min per session 	12 weeks	ECG, Holter monitoring, echocardiography, graded maximal exercise test	<ul style="list-style-type: none"> • Decrease in resting blood pressure and increase in peak oxygen consumption and treadmill time
Brassard [67]	7	History of Fontan procedure	11–26	<ul style="list-style-type: none"> • 3 20–30-min sessions 3 times per week • Aerobic and resistance training 	8 weeks	Pulmonary function evaluation, neuromuscular function evaluated via ergoreceptor activity monitoring, skeletal muscle strength, endurance evaluation, exercise testing	<ul style="list-style-type: none"> • Reduction in ergoflex contribution to absolute values of systolic blood pressure (suggested improvement in skeletal muscle function)
Dedieu [68]	33	CHD	8–40	<ul style="list-style-type: none"> • 2 days a week training 	8 weeks	Exercise testing	<ul style="list-style-type: none"> • Increase in maximum heart rate, exercise duration and metabolic equivalents, improvement in quality of life
Duppen [69–72]	93	Tetralogy of Fallot and history of Fontan procedure (vs. healthy controls)	10–25	<ul style="list-style-type: none"> • 3 1-h sessions per week • Sports activities twice per week • 10-min warm-up • 40-min aerobic training (brisk walking, dynamic play, jogging/running/-bicycle) • 10-min cool down • Heart rate monitor given workload adjusted to heart rate levels • Control: life as usual 	12 weeks	Exercise testing, psychological assessment, semi-structured interview, Web-based health-related quality of life assessment	<p>All ages:</p> <ul style="list-style-type: none"> • Peak load increased in the intervention group • Ventricular systolic parameters did not change in the intervention group • No adverse events <p>Ages 10–15:</p> <ul style="list-style-type: none"> • Improved cognitive functioning • Self-report of improved motor functioning after the sports-related intervention • Parent report of improved social functioning

Table 3 (continued)

References	N	Patient population	Age (in years)	Program protocol	Program duration	Measures	Outcome/results
							Ages 16–25: • No differences in quality of life
Frederiksen [73]	129	CHDs	10–16	• 2-week intensive rehabilitation at sports center or twice per week program	2 weeks or 5 months	Exercise testing	• Improvement in uptake of peak level of oxygen • Increase in physical activity • Decrease in internalized distress
Opocher [74]	10	History of Fontan procedure	7–12	• Twice per week lessons for 3 months • Once per week lessons for 4 months • At-home training twice per week for 30–45 min	8 months	Exercise testing	• Improved aerobic capacity • Increase in maximal oxygen consumption • Decrease in heart rate curve • Increased oxygen pulse curve during submaximal exercise
Rhodes [62•, 75, 76]	30	CHDs	8–17	• 2 1-h sessions per week • Aerobic and weight resistance exercises • Recommended home program of twice per week	12 weeks	Exercise testing	• Improved peak oxygen consumption, peak work rate, ventilatory anaerobic threshold, 1-min recovery rate • After 1 year, peak oxygen consumption, work rate, and 1-min heart rate recovery remained improved
Sklansky [7]	11	History of tetralogy of Fallot repair	N/A	• 3 30-min sessions per week • Use of cycle ergometer	8 weeks	Exercise testing	• Decrease in submaximal heart rate, submaximal cardiac output, maximal oxygen uptake, respiratory rate, and maximal treadmill time
Wittekind [78•]	10	History of Fontan procedure	8–15	• 2 1-h sessions per week • 5- to 10-min warm-up and stretching • 30-min aerobic training • 15-min low-resistance and high-repetition strength training • 5-min cool down	12 weeks	Cardiopulmonary exercise testing, ECHO, heart rate, blood pressure, VO ₂ , VCO ₂ , minute ventilation, perceived exertion using the Borg	• Increase in peak indexed oxygen consumption increased, peak oxygen pulse, and indexed oxygen consumption at ventilator anaerobic threshold
Home-based Rehabilitation							
Amiard [79•]	23	CHDs with history of surgical repair	15 ± 1.4	• 45-min sessions consisting of 10-min warm-up and 5-min stretching, and use of a home cycle ergometer	8 weeks	Exercise testing	• No strong improvement in aerobic capacity or ventilatory threshold
Hedlund [80]	55	History of Fontan procedure (vs. healthy controls)	Mean 14.2	• Individualized endurance training program. • 2 45-min endurance training sessions per week	12 weeks	6-min walk test, ergometer cycle test, and Pediatric Quality of Life Inventory	After intervention: • Increase in average intensity on the Borg scale for activities • Increase in 5-min walk test

Table 3 (continued)

References	N	Patient population	Age (in years)	Program protocol	Program duration	Measures	Outcome/results
				<ul style="list-style-type: none"> Activities included running, skiing, jogging, skiing, cycling, swimming, riding, dancing, football 			<ul style="list-style-type: none"> VO₂ max did not increase Quality of life improved At 1-year follow up Borg intensity back to average 6-min walk test remained improved Quality of life was sustained
Jacobsen [81•]	13	History of Fontan procedure	8–12	<ul style="list-style-type: none"> 45-min routine with dynamic and static exercises on video and handout 3–4 times per week 3 in-person sessions at 0-week, 6-week, and 12-week marks Use of Fitbit Flex to monitor adherence 	12 weeks	VO ₂ max estimates, 20-m shuttle test run, baseline height, weight, body mass index, blood pressure, heart rate, ECHO Primary outcome: physical function and quality of life Secondary outcome: change in exercise capacity	<ul style="list-style-type: none"> 93% program completion Improvements in VO₂ max, quality of life, and exercise capacity
Longmuir [82]	61	History of Fontan procedure	5.9–11.7	<ul style="list-style-type: none"> 2 models with a focus on increasing physical activity by play. Both models included specific daily activities, were parent-led, and had at-home and community-based design 1-year commitment to 2 activity hours per week 	2 years	Tests of gross motor development, aerobic step test, grip strength, hamstring flexibility, body mass index, health-related fitness, exercise testing, and activity attitudes assessment	<ul style="list-style-type: none"> No difference was noted in secondary outcomes (VO₂ max, exercise fitness scores, etc.) Improved gross motor skills
Moalla [83, 84]	31	CHDs in New York Heart Association Class II or III (vs. healthy controls)	12–16	<ul style="list-style-type: none"> 2 groups with CHDs: untrained and control. 3 sessions per week 10-min warm-up 10-min work and 5-min recovery intervals on a stationary bike 5-min cool down Intensity set a heart rate of ventilatory threshold obtained during cardiopulmonary testing 	12 weeks	Pulmonary function tests, exercise testing, and near-infrared spectroscopy assessment used to evaluate oxygenation of respiratory muscles	<ul style="list-style-type: none"> Improved walking distance Power output, VO₂, heart rate, and ventilation increased slightly at peak exercise and significantly at ventilatory threshold level
Minamisawa [85]	16	History of Fontan procedure	11–25	<ul style="list-style-type: none"> Sessions 2–3 days per week 5-min warm-up with 20–30 min jogging to target heart rate 	8–12 weeks	Exercise testing	<ul style="list-style-type: none"> Improved peak oxygen consumption and workload
Morrison [86]	143	CHDs	12–20	<ul style="list-style-type: none"> 1-day structured group activity 6 months of activity based on written exercise plans 	6 months	Exercise testing	<ul style="list-style-type: none"> Improved peak oxygen consumption Increased moderate to vigorous physical activity

Table 3 (continued)

References	N	Patient population	Age (in years)	Program protocol	Program duration	Measures	Outcome/results
Stieber [87]	20	Single ventricle physiology, transposition of great arteries, history of superior c avopulmonary connection procedure or arterial switch operation	1–2	<ul style="list-style-type: none"> • 5 2-week sessions • Play activities for a total of 10 min or more each day with a goal of 20 min total (10 min per development goal) 	10 weeks	Peabody Developmental Motor Scale, 2nd Edition	<ul style="list-style-type: none"> • Expected rate of motor development was achieved in both groups
Patel [88]	11	History of heart transplant	Mean 14.7	<ul style="list-style-type: none"> • Aerobic exercise 3 days per week • Strength training 2 days per week 	12 weeks	Exercise testing	<ul style="list-style-type: none"> • Increase in peak oxygen consumption, endurance time, strength measurements

* Exercise testing typically included the following measures: electrocardiogram (ECG), blood pressure, maximal oxygen consumption (VO_2), work performed, maximal oxygen pulse, maximum including oxygen uptake, carbon dioxide production (VCO_2), minute ventilation, maximal ventilator, and metabolic equivalents heart rate

Below, we describe pediatric cardiac rehabilitation programs designed for inpatient and outpatient settings as well as home-based training. Across the different settings, programs typically adopted a team approach with physicians, rehabilitation therapists, and other health providers. Indeed, it has previously been recommended that programs include dietitians, physical therapists, and occupational therapists at minimum and be coordinated by a pediatric cardiologist and a pediatric physiatrist [30•]. Staff should be trained to handle medical emergencies, and the staff-to-patient ratio should not exceed 1 to 4 at any time, per prior recommendations [30•].

Inpatient Acute Care Rehabilitation

The literature is limited in regard to programs that include rehabilitation during the acute care hospitalization period. Still, we identified two reports describing inpatient acute care rehabilitation, one featuring patients with a ventricular assist device and one focused on patients hospitalized for heart failure [65, 66]. The programs demonstrated the safety and feasibility of an acute care model both in the intensive care setting and on a standard hospital floor, with no adverse events related to physical activity. One of the rehabilitation programs additionally documented that patients were able to achieve the therapeutic goals established for them [65].

Outpatient Rehabilitation

We identified nine outpatient programs, described across 14 studies [61, 62•, 67–77, 78•]. The programs included patients as young as 8 years old; although three of the programs included young adults and one included patients as old as 40 years, they all enrolled children and/or adolescents. All of the studies

evaluated effects of exercise in patients with CHDs. The duration of the programs ranged from 2 weeks to 8 months, with structured activity typically two to three times per week, typically for 30 min to 60 min per session. The programs all included some form of warm-up and cool-down periods (typically 5 min to 10 min each). All of the programs included an aerobic component to the exercise training program, and some also included strength training. All of the programs demonstrated an improvement in maximal oxygen output. Studies also reported exercise duration, metabolic equivalents, and improved quality of life. One of the programs also documented self-reported improvements in cognitive functioning and parent-reported improvements in social functioning [69–72].

Home-Based Rehabilitation

Nine programs, summarized across ten reports, described home-based rehabilitation programs [79•, 80, 81•, 82–88]. Although the majority of studies focused on school-age and adolescent patients with CHDs, there was one program that focused on toddlers [87], one that included young adults [85], and one that enrolled patients following heart transplantation [88]. Programs often required 8 weeks to 12 weeks of home-based exercise and included initial and follow-up sessions in person with health providers, so as to provide education on the program and monitor progress. Progress could also be monitored with digital tools, such as heart rate monitors, accelerometers, and Fitbits. Echoing the design of outpatient programs, home-based rehabilitation programs typically involved sessions of an hour or less with 5-min to 10-min warm-up and cool-down periods and focus on aerobic exercise. Although training was often associated with improved exercise capacity, better cardiorespiratory outcomes, and improved quality of

life and psychosocial functioning, one study did not find significant changes in patients after the intervention [79•]. Interestingly, the program with toddlers reported improvements in gross motor functioning, daily physical activity, and family life up to 2 years later [87], and a program with school-age children documented improvements in exercise capacity and quality of life up to a year after the intervention [80]. None of the programs reported significant adverse events.

Cognitive Rehabilitation

The vast majority of rehabilitation programs among children with cardiac conditions have focused on exercise training, as a function of children's complex medical needs. However, we mention one study underway to examine cognitive rehabilitation among children with cardiac conditions. Newburger [89] is investigating the effects of a computer-based training program to enhance the executive functioning skills of children with CHDs.

Current Directions at Our Institution

At the UPMC Children's Hospital of Pittsburgh (CHP), we developed a cardiac rehabilitation program modeled after recommendations from previously developed programs, as described above. The program seeks to establish a reproducible and sustainable protocol for children with pediatric-onset or congenital cardiac disease, in an effort to systematically influence and mitigate the effects on physical health, neurodevelopmental outcomes, and quality of life. Specifically, the goals of the program are to (1) improve exercise function and capacity; (2) improve heart rate recovery and response to exercise; (3) decrease the effects of deconditioning; (4) improve physical function and age appropriate level of play and adaptive living; (5) improve patient and family confidence in exercise and participation in age appropriate activities; (6) promote global wellness, including an active lifestyle and healthy nutrition choices; and (7) improve overall adjustment and quality of life post transplantation.

With this framework in mind, we established multidisciplinary inpatient and outpatient cardiac rehabilitations programs, which involve medical evaluation, prescribed therapies, nutritional and physical education geared toward improving the physiologic effects of long-term cardiac illness, and consultation for developmental, neuropsychological, and psychological services. The team, headed by a pediatric cardiologist and pediatric physiatrist, also includes physical therapists, occupational therapists, speech and language pathologists, dietitians, neuropsychologists, and pediatric psychologists. This team meets at a regularly scheduled time to review patients for clinic care and to advance the cardiac rehabilitation program's development.

Currently, our institution is targeting children on the pathway to heart transplantation for participation in pediatric cardiac rehabilitation; however, we hope to expand our program more broadly to children with CHDs and cardiomyopathies not requiring heart transplantation. The program structure includes providing a multidisciplinary evaluation prior to transplantation, including assessments from medical and therapy providers. Based on the evaluation, the team provides a cohesive set of recommendations, including a potential course of therapies prior to transplantation. Following cardiac transplantation, patients are typically engaged in a 12-week outpatient course of physical therapy three times per week (with sessions lasting between 30 and 60 min), with aerobic conditioning, resistance, and strength training. Sessions include warm-up and cool-down periods. As needed, patients are engaged in occupational therapy with a focus on adaptive skills following surgery, speech and language therapy with a focus on cognitive restructuring, and nutrition and physical activity education. These sessions can occur at facilities associated with the hospital for families that live close by; otherwise, a prescription detailing needed interventions is provided to the family to take to a facility in their community. As needed, patients are engaged in psychotherapy to aid with their psychosocial adjustment. Patients may also be referred for neuropsychological testing to determine their cognitive needs, as they return to the school and home environment; evaluations are conducted following guidelines for assessment in children with cardiac disorders [90•].

Conclusion

Children can benefit from pediatric cardiac rehabilitation programs from physical health and psychosocial perspectives, and more importantly, these programs can be tolerated with minimal adverse events. Although center-based programs may allow for a greater degree of refinement in individual patient goals and monitoring of their health status, the limitation of proximity to a tertiary care institution is not necessarily a barrier (particularly among children with chronic concerns, such as CHDs), as demonstrated by the efficacy of home-based programs. The home-based program has the potential to limit the burden of transportation and potentially time off work for parents who bring their child to a facility for training. Still, home-based programs should continue to be monitored by both a cardiologist and a pediatric physiatrist. Further research is needed to demonstrate sustained long-term benefits of interventions, to understand the impact on quality of life for the family system, and to explore long-term impact of cognitive interventions within pediatric cardiac rehabilitation programs.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflicts of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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