Exercise training in paediatric congenital heart disease: fit for purpose?

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ABSTRACT

Exercise and physical activity (PA) have been shown to be effective, safe and feasible in both healthy children and children with congenital heart disease (CHD). However, implementing exercise training as an intervention is still not routine in children with CHD despite considerable evidence of health benefits and well-being. Understanding how children with CHD can safely participate in exercise can boost participation in PA and subsequently reduce inactivity-related diseases. Home-based exercise intervention, with the use of personal wearable activity trackers, and high-intensity interval training have been beneficial in adults' cardiac rehabilitation programmes. However, these remain underutilised in paediatric care. Therefore, the aims of this narrative review were to synthesise prescribed exercise interventions in children with CHD, identify possible limitation to exercise training prescription and provide an overview on how to best integrate exercise intervention effectively for this population into daily practice.

INTRODUCTION

Congenital heart disease (CHD) is the most common birth defect, affecting an estimated 130 million newborns yearly,[1] has a prevalence of 9.4 per 1000 live births[2] and is a leading cause of infant death.[3] The aetiology of CHD is multifactorial, with genetic, teratogenic exposure and maternal diabetes being attributed to the incidence of CHD.[2] Major advances in the field of paediatric cardiology and surgery over the decades have dramatically improved the survival of infants born with CHD. Currently over 90% children born with CHD have the prospect of surviving into adulthood.[4]

Exercise training is a subcomponent of physical activity (PA) that systematically aims to improve specific physical fitness components, namely cardiorespiratory fitness, muscular strength, body composition and flexibility, and relates to the ability to perform any PA.[5] Exercise and PA have been shown to maintain and improve health and quality of life (QoL) and reduce all-cause mortality in healthy children as well as those with CHD.[3] Moreover, the emergence of COVID-19 has highlighted the benefits of home-based exercise intervention (HBEI) as a rehabilitation modality.[6 7] Therefore, this narrative review aims to synthesise prescribed exercise interventions in children with CHD, identify possible limitations to exercise prescription and provide an overview on how to best use exercise interventions in this patient group.

METHODS

A systematic approach was conducted for this narrative review using the electronic databases PubMed, Scopus and Google Scholar up to June 2021. The following search terms were used: 'congenital heart disease', 'exercise training', 'exercise programme', 'physical activity', 'home-based', 'centre-based', 'hospital-based', 'high-intensity interval training', 'HIIT', 'HIIE', 'wearable devices', 'activity tracker' and 'physical activity monitor'. Studies were included if they were a randomised clinical trial or a systematic review or if they contained structured cardiac rehabilitation programme with an exercise training component in cardiac patients. The population included children and young people (age up to 18 years) with any form of CHD.

PA IN CHILDREN AND YOUNG PEOPLE

The prevalence of physical inactivity has become a global health crisis, responsible for an estimated five million deaths worldwide.[8] Approximately 20 million adults in the UK were physically inactive, predisposing them to a greater risk of heart and circulatory diseases,[8] diabetes,[9] cancers,[10] and ultimately premature death.[8] Almost 60% of adults in the UK are unaware of the PA guidelines and only 47% of UK children meet the current PA guidelines.[11] This decline in PA has been anticipated to cause an additional health economic burden of £1.2 billion annually to the English National Health Service.[8]

Although cardiovascular disease (CVD) becomes most evident in adulthood, the development of the disease begins in childhood and in young people,[12] with physical inactivity and poor diet being major modifiable factors of atherosclerosis predisposition,[13] compounded by excessive screen time and associations to sedentary living and less time spent in play.[12] Early preventative and/or intervention through PA should begin as early as possible in childhood to instil habitual behaviours and improve cardiovascular health markers.[14] How inactivity currently affects young people with CHD is poorly understood. However, there are some data that showed children with CHD have reduced activity levels compared with healthy peers[15 16] and are at increased risk of acquired CVD. Currently, approximately 80% of all young adults with CHD have at least one cardiovascular risk factor.[17]

The benefits of regular PA and exercise training in improving health have long been established with a strong inverse relationship with morbidity and strong positive relationship with mortality.[18 19]

Table 1 Evidence on the relationship between physical activity and health risk factors in healthy children and adolescents

Study	Age range	Population	Primary objectives	Assessment method	Findings
Kong *et al*[100]	6–20 years n=1882 (780 boys)	Healthy	To examine the cross-sectional association of self-reported level of physical activity and cardiovascular risk factor in Hong Kong Chinese youth.	CHUK-PARCY Questionnaire. Cardiovascular risks (waist circumference, blood pressure, fasting plasma glucose, lipid).	In this cohort, 21.5% reported high level of PA, with boys being more active than girls (32.1% vs 14.1%, p<0.001). As for final regression analysis, girls and high PA were negatively associated while increase in PA remained independently associated with low-risk score for cardiovascular risk.
Janz *et al*[101]	5–17 years n=530 (267 boys)	Healthy	To examine the developmental trajectories of objectively measured PA from childhood to adolescence to discern if MVPA predicts bone strength in healthy children.	Activity monitor (Actigraph): 4 days (ages 5 and 8 years); 5 days (ages 11, 13, 15 and 17 years). DEXA.	Participants who developed the most MVPA gained greater bone mass and better geometry at 17 years when compared with less active peers. Higher level of MVPA during childhood positively associated with bone strength in late adolescence even after drastic reduction in PA during puberty.
Carson *et al*[102]	9–15 years n=315 (128 boys)	Healthy	To examine the longitudinal associations between different PA intensities and cardiometabolic risk factors among a sample of Canadian youth.	BMI z-score. Waist circumference. Cardiorespiratory fitness (VO_2 max). Blood pressure (SBP).	After 2 years of follow-up, VO_2 max increased (p<0.01) and waist circumference decreased (p<0.04, boys only) in a dose–response manner across baseline to vigorous-intensity PA. SBP reportedly decreased from 121.8 to 115.3. BMI z-score at follow-up and conditional BMI z-score were significantly lower after vigorous PA. These findings suggest vigorous-intensity PA may reduce the risk of cardiometabolic disease among youth.
Barker *et al*[103]	12.5–17.5 years n=534 (252 boys)	Healthy	To examine the independent association between PA intensities, sedentary time (ST), television viewing, cardiorespiratory fitness (CRF) and muscular fitness (MF) with cardiovascular disease (CVD) risk in youth.	Activity monitor (Actigraph GT1M). Self-reported sedentary behaviour questionnaire. 20 m shuttle run. Hand dynamometer.	LPA had a significantly positive independent relationship (p=0.046) while VPA had a negative independent relationship (p=0.01) with clustered CVD risk. MPA did not consistently demonstrate as strong significant predictor of CVD risk factors. ST was not significantly related to CVD risk factors. Increased television viewing time was an independent predictor of elevated CVD risks (p=0.019), TG, TG:HDL-C and HOMA-IR. Both CRF (p<0.002) and MF (p<0.009) were negatively related with body composition indices and clustered CVD risk.
Janssen *et al*[104]	7–15 years n=502 (7 years) n=506 (9 years) n=420 (12 years) n=306 (15 years) (almost equal split between boys and girls)	Healthy	To examine non-linear longitudinal associations between MVPA and adiposity by weight status across childhood and adolescence.	Activity monitor (Actigraph GT1M), 7 days except water activity.	Higher levels of MVPA are associated with lower levels of adiposity during childhood and adolescence. Change in MVPA and change in BMI and FMI were stronger in those with higher BMI and FMI (eg, 1 hour/day more MVPA was associated with 1.5 kg/m^2 and 2.7 kg/m^2 lower FMI at the 50th and 90th FMI percentiles, respectively).
Wellman *et al*[105]	12–17 years n=993 (481 boys)	Healthy	To examine the relationship between PA intensity and frequency and the likelihood of having high blood pressure in a population-based cohort of adolescents from Montreal, Canada.	Self-reported questionnaire. Blood pressure (SBP and DBP).	Blood pressure level was categorised as normal/elevated/hypertensive. Engaging in any level of PA especially in more intense than light over the past year is associated with lower chances of having blood pressure in the hypertensive range, and suggests that PA may protect against high blood pressure in adolescents (OR 0.93 (95% CI 0.88 to 0.97); OR 0.97 (95% CI 0.94 to 0.99)).

BMI, body mass index; CHUK-PARCY, The Chinese University of Hong Kong-Physical Activity Rating for Children and Youth; DBP, diastolic blood pressure; DEXA, dual-energy X-ray absorptiometry; FMI, fat mass index; HOMA-IR, Homeostasis Model Assessment of Insulin Resistance; LPA, low physical activity; MPA, moderate physical activity; MVPA, moderate-to-vigorous physical activity; PA, physical activity; SBP, systolic blood pressure; TG, triglycerides; TG:HDL-C, triglycerides to high-density lipoprotein-cholesterol ratio; VO_2max, maximum oxygen uptake; VPA, vigorous physical activity.

Evidence on the relationship between PA and health risk factors during childhood and adolescent years is summarised in table 1. PA has been shown to improve aerobic fitness[20 21] and reduce CVD risk factors[22 23] through moderate-intensity continuous training (MICT) or high-intensity interval training (HIIT). Also, supramaximal HIIT performed at a higher intensity than conventional HIIT was found to be well tolerated and feasible in adolescent boys.[24] However, its effect on other health-related risk factors is equivocal (see table 2).

The latest WHO Physical Activity Guidelines (2020) advocate children and adolescents from 5 to 17 years old should engage in an average of 60 min per day of moderate-to-vigorous PA, mostly aerobic activity, across the week, and vigorous PA including bone and muscle strengthening at least three times a week, and minimise sedentary activities.[25] Only 21% of boys and 16% of girls from 5 to 15 years of age in the UK achieved these PA guidelines in 2015.[26] Consequently, physical inactivity contributed to almost one in ten premature deaths through inactivity-related diseases, for example, coronary atherosclerosis.[27] This led to an increased awareness among paediatric health professionals of sedentary lifestyle-associated morbidity among children and adolescents,[28] particularly in children with CHD. There is accumulating evidence on the benefits of regular exercise for children with CHD,[29–47] demonstrating PA can be functionally beneficial and consequently improve overall health. However, modifications may be necessary when prescribing exercise intensity and volume for children with CHD specific to individual anatomical lesion and functional status.[48 49]

EXERCISE TRAINING IN CHD

Exercise training interventions have been used successfully in the rehabilitation of adults with CHD[50–57]; however, it is less established among children and young people.[58] In a recent survey among paediatric cardiac clinic staff across the UK, 36% of clinicians reported that written advice about exercise is never provided despite being discussed during consultation.[59] Less than half of all clinicians surveyed failed to consider published guidelines when discussing exercise with patients.[59] This failure to consider published guidelines is a concern because inactivity from the early age would have an impact on basic motor development and QoL.[46] A small proportion of paediatricians and

Table 2 Summary effects of exercise intervention in children and adolescents (healthy and specific population)

Study	Age range	Population	Primary objectives	Assessment method	Findings
Nourry *et al*[106]	8–10 years n=18 (11 boys)	Healthy prepubescent	To investigate the effects of short-duration running training (HIIT) on resting and exercise lung function in healthy prepubescent children.	FVC and FEV_1. PEF. MEF. Peak VO_2.	Significant increase in lung function capacity (FVC, FEV_1, PEF, MEF) was found after 8 weeks of HIIT in the training group. Cardiorespiratory function (peak VO_2, VE) was also reported to significantly increase. Overall, HIIT running improves resting lung function and leads to greater exercise ventilation, reflecting better effectiveness in lung function among prepubescent children.
Tjønna *et al*[22]	14 years n=54 (26 boys)	Obese	To compare the effects of multidisciplinary approach (MA) and aerobic interval training (AIT) on cardiovascular risk factors in overweight adolescents.	BMI. DEXA. SBP. DBP. VO_2 max. 1-RM leg test.	The AIT group successfully improved VO_2 max (p<0.01) and endothelial function (SBP and DBP) and favourable in reducing BMI, fat percentage and mean arterial BP following 3 months and 12 months of follow-up among overweight adolescents.
Baquet *et al*[20]	8–11 years n=60 (23 boys)	Healthy	To show if the use of continuous-running training vs intermittent-running training has comparable or distinct impact on aerobic fitness in children.	Cardiovascular fitness (peak VO_2). Maximum aerobic velocity.	Both continuous-running and intermittent-running training improve peak VO_2 (+7% CTG; +4.8% ITG) and MAV (+8.7% CTG; +6.4% ITG).
Lambrick *et al*[21]	8–10 years n=55 (32 boys)	Healthy normal weight vs obese	To assess the effectiveness of a 6-week HIIT, child-specific game intervention in improving physiological and anthropometrical indices of health and fitness in children of different body mass between normal and obese.	VO_2 max. Peak running speed. Anthropometrical data. Submaximal testing.	The HIIT group demonstrated improvement in VO_2 max and peak running speed and reduction in oxygen cost of submaximal exercise test (p<0.05). A decrease in waist circumference and an increase in muscle mass were observed between assessments in obese patients (both p<0.05).
Racil *et al*[23]	13–15 years n=47 girls	Obese	To examine the effects of high-intensity (HIIT) vs moderate-intensity (MIIT) interval training on cardiovascular fitness, leptin levels and RPE in obese female adolescents.	Max HR. SBP/DBP. Blood glucose. Blood leptin. RPE index.	Both MIIT and HIIT showed improvement in BMI and fat percentage. Only HIIT showed reduction in waist circumference. VO_2 max was reported to be increased in both training groups, including decrease in rate pressure product due to positive change in BP and HR. Blood leptin, blood glucose and RPE index were significantly lowered in both training groups. HIIT was suggested to induce more positive effects on health determinants.
Chuensiri *et al*[24]	8–12 years n=48 boys	Obese	To determine whether HIIT and supramaximal high-intensity intermittent training (supra-HIIT) would improve vascular structure and function, BMR, physical fitness and CVD traditional risk factors in obese preadolescent boys.	BC and WHR. HR, BP, VO_2 peak, work rate and leg muscle strength. Arterial stiffness and thickness. physical activity enjoyment score (PACES). Physical activity (pedometer). Blood serum analysis.	Both HIIT and supra-HIIT did not affect body mass, body fat percentage and waist circumference. Peak VO_2 increased in both HIIT and supra-HIIT (p<0.05). Increase in resting metabolic rate was observed in both intervention groups than the control (p<0.05). Arterial stiffness and cIMT decreased after 12 weeks in the intervention groups (all p<0.05). FMD reportedly increased in both HIIT and supra-HIIT groups (all p<0.05). Both HIIT and supra-HIIT are effective and time-efficient lifestyle modification strategies for obese preadolescent boys.
Ingul *et al*[107]	6–17 years n=99	Obese	To compare the effects of HIIT, MICT and nutrition advice intervention on resting LV peak systolic tissue velocity (S′) in obese children.	Cardiac function and structure. Vascular function. Cardiorespiratory fitness.	12 weeks of HIIT and MICT were equally efficacious and superior to nutrition advice, in terms of normalising resting LV S′ in obese children (estimated mean difference=1.0 cm/s, 95% CI 0.5 to 1.6 cm/s, p<0.001; estimated mean difference 0.7 cm/s, 95% CI 0.2 to 1.3 cm/s, p=0.010, respectively).

BC, body composition; BMI, body mass index; BMR, basal metabolic rate; BP, blood pressure; cIMT, carotid intima-media thickness; CTG, continuous-training group; CVD, cardiovascular disease; DBP, diastolic blood pressure; DEXA, dual-energy X-ray absorptiometry; FEV_1, forced expiratory volume in 1 s; FMD, flow mediated dilation; FVC, forced vital capacity; HIIT, high-intensity interval training; HR, heart rate; ITG, interval-training group; LV, left ventricle; MAV, maximum aerobic velocity; Max HR, maximum heart rate; MEF, maximal expiratory flow; MICT, moderate-intensity continuous training; peak VO_2, peak oxygen consumption; PEF, peak expiratory flow; 1-RM, 1-repetition maximum; RPE, ratings of perceived exertion; SBP, systolic blood pressure; VE, minute ventilation; VO_2max, maximum oxygen uptake; WHR, waist to hip ratio.

nurses cited the risk associated with PA as a barrier, whereas the majority of clinicians do not have the time, resources and specific knowledge to confidently provide exercise advice for youngsters with CHD.[59] Nevertheless, physiological mechanisms in response to exercise can vary widely across different CHD types[47 60 61] and are not fully understood, which underline the need for more physiological studies among paediatric patients.[49]

Several studies investigating exercise in children with CHD have led to convincing evidence on exercise capacity and QoL benefits[31 32 35] and viewed it as a safe[36 58 62 63] and cost-effective[64] intervention alongside CHD care. However, the effectiveness of exercise interventions remains ambiguous, with limitations confounding the research findings, for example, small sample size,[36 41] absence of healthy controls,[33 34 40] insufficient exercise volume,[35 36 38] patient psychosocial factors[12] or heterogeneous cohorts.[45]

LIMITATIONS OF PREVIOUSLY PRESCRIBED EXERCISE INTERVENTIONS

Understanding why previous studies of exercise training have resulted in little or no improvement in exercise capacity and health-related components might help to improve future strategies.[65] Limitations of available studies relate to dosage, sample size, absence of healthy group, HIIT versus MICT, home versus hospital-based, and monitoring exercise.

Exercise dosage, sample size and healthy control group

Exercise dosage is the product of FITT (frequency, intensity, time and type of exercise) and the application of training principles.[66] When prescribing exercise, dosage is critical to maximise intervention efficiency, but many previous studies did not offer accurate dosage information.[30 35 36 38] In one study among children with Fontan circulation, there was no significant improvement in maximal exercise capacity (Maximum oxygen uptake, VO_2 max baseline 35.0±5.1 mL/kg/min vs postexercise 35.6±6.3 mL/kg/min).[35] The exercise intensities were low (Borg Scale (6–20) baseline 13±2 vs postexercise 14±0), as patients were instructed not to exercise at maximal effort, and this factor probably contributed to the negative outcome.[35] In another study, the dyspnoea-based intensity did not significantly improve exercise capacity in children.[38] Exercise selection based on participants' preferences (ie, cycling, jogging, football) is encouraged for as long as exercise is performed at sufficient intensities.[35] This limitation in exercise dosage can be addressed by following the recommendation by Budts *et al*[49] to

Table 3 Home-based exercise intervention evidence among CHD populations (children and young adults)

Authors	Patients with CHD	Age (years)	Study design	Duration of programme	Duration/frequency/ intensity	Description of training	Monitoring	Outcomes measured	Results
Minamisawa *et al*[44]	History of Fontan procedure	11–25	Quasi-experimental	8–12 weeks	5 min warm-up with 20–30 min jogging to target HR. 2–3×/week.	Fast walking/jogging.	Phone calls every 2–3 weeks to ensure compliance and safety.	Peak VO_2, work rate HR max, SaO_2, VE.	Improved peak VO_2 and work rate.
Moalla *et al*[43]	Class II and III NYHA	12–15	RCT	12 weeks (individualised)	10 min × 5 min rest (repeat until 45 min). 3×/week. HR corresponding to the ventilatory threshold.	Interval exercise, cycle ergometer.	HR monitor.	6 min walk test, peak work rate, VO_2 max, HR max, SaO_2 VE max.	Improvement in WD post-exercise. A slight increase in peak work rate, VO_2 max and HR max at ventilatory threshold compared with maximum exercise, and significant relationship between WD and VO_2 max.
Moalla *et al*[41 42]	Class II and III NYHA	12–15	RCT	12 weeks (individualised)	10 min × 5 min rest (repeat until 45 min). 3×/week. HR corresponding to the ventilatory threshold.	Interval exercise, cycle ergometer.	HR monitor.	FVC, FEV_1, TLC, NIRS, peak work rate, VO_2 max, SBP, DBP, VE, HR, MVC, MVC 50% time to exhaustion.	Improvement in cardiorespiratory performance at submaximal intensity. A respiratory oxygenation improvement during exercise associated with improvement in exercise tolerance in TG after training among patients with CHD. Improvement in muscle maximum voluntary contraction and muscular endurance and increased oxygenation and muscle recovery.
Amiard *et al*[38]	CHD with history of surgical repair	15±1.4	RCT	8 weeks	10 min × 5 min rest (repeat until 45 min). 3×/week. HR corresponding to the dyspnoea threshold ±5 beats/min.	Interval exercise, cycle ergometer.	HR monitor.	Power output, VO_2 max, HR max, VE max.	No strong improvement in aerobic capacity or ventilatory threshold.
Stieber *et al*[46]	ASO for tGA and SCPC for palliation of the functional single ventricle	1–2	Quasi-experimental	10 weeks	10 min or more each day (goal 20 min total, 10 min per development goal). 7×/week.	Parent-led, play-based activities.	Parents were contacted biweekly for progress, feedback and new activities; also served as reminder to implement intervention.	Peabody Developmental Motor Scale Second Edition.	Expected rate of motor development was achieved in both ASO and SCPC.
Morrison *et al*[45]	Various types of CHD	12–20	RCT	1–6 months (individualised)	6 activity intervention days. Motivational interviewing techniques promoting exercise/activity. Additional training plan to implement at home (verbal).	Motivational interview to promote exercise.	Participants were contacted once a month for progress/problem.	Predicted maximum VO_2, PA.	Increase in predicted maximum VO_2 and improvement in time spent for PA postintervention.

Continued

Table 3 Continued

Authors	Patients with CHD	Age (years)	Study design	Duration of programme	Duration/frequency/ intensity	Description of training	Monitoring	Outcomes measured	Results
Longmuir *et al*[40]	History of Fontan procedure	5.9–11.7	RCT	2 years	1.5–2 hours/week. 4×/week. No specified intensity.	Two intervention models with the aim to increase PA by play. Both models were parent-led, home/ community- based and had included specific daily activities.	Participants were contacted once a month (mid-month) for feedback and to encourage compliance.	Test of gross motor development, aerobic step test, grip strength, hamstring flexibility, BMI, health-related fitness, exercise testing and activity attitudes test.	Improvement in gross motor skills associated with increase in MVPA. No difference in secondary outcomes (VO_2 max, exercise fitness scores, grip strength, flexibility, BMI percentile and child's self-reported adequacy and predilection for PA).
Klausen *et al*[108]	Complex CHD	13–16	Randomised clinical trial	52 weeks (individualised)	*Modality:* text/technology-based PA encouragement. *Intensity:* text encouraged 'high intensity', but it is probably more appropriate to state MVPA due to a loose definition of high intensity.	Activity encouragement via text messages.	Self-reported adherence using an app.	Peak VO_2, PA (accelerometer)	No significant improvement in peak VO_2 and no improvement in time spent for PA post 52 weeks activity encouragement.
Jacobsen *et al*[36]	Fontan	8–12	–	12 weeks (standardised)	45 min/session. 3–4×/week. No specified intensity.	DVD home workout. Extra PA outside programme not restricted.	Fitbit, phone call, daily activity journal.	20 m shuttle run test, HRQoL of parents and patients (CPET, ECG, ECHO recorded but not reported).	Improvement in 20 m shuttle distance and estimated VO_2 max.
Hedlund *et al*[35]	Fontan	12–25	–	12 weeks (endurance)	45 min/session. 2×/week. Borg Scale 6–20 ('not to exert maximal effort').	Chosen sports with instructor-led activities near home or schools.	Instructor-monitor. Intensity/duration recorded and reported weekly.	VO_2 max, HR max, BP max, work rate max, 6 min walk test, QoL questionnaires.	Improved HRQoL in physical and psychological domains after 12 weeks for the intervention group.

ASO, arterial switch operation; BMI, body mass index; BP, blood pressure; BP max, maximum blood pressure; CHD, congenital heart disease; CPET, cardiopulmonary exercise test; DBP, diastolic blood pressure; ECHO, echocardiogram; FEV_1, forced expiratory volume in 1 s; FVC, forced vital capacity; HR, heart rate; HR max, heart rate maximum; HRQoL, health-related quality of life; MVC, maximum voluntary contraction; MVC 50%, muscular endurance at 50% time to exhaustion; MVPA, moderate-to-vigorous physical activity; NIRS, near-infrared spectroscopic; NYHA, New York Heart Association; PA, physical activity; peak VO_2, peak oxygen consumption; QoL, quality of life; RCT, randomised controlled trial; SaO_2, oxygen saturation; SBP, systolic blood pressure; SCPC, superior cavopulmonary connection; TG, training group; tGA, transposition of the great arteries; TLC, total lung capacity; VE, minute ventilation; VE max, maximum minute ventilation; WD, walking distance.

Table 4 Registered clinical trials using physical activity and/or exercise as intervention in children and adolescents with CHD (2017–2021)

Registry	Trial registration number	Study title	Intervention	Status	Last update posted
ISRCTN registry	ISRCTN16613503	Physical activity and exercise pathway for patients with congenital heart disease	Home-based exercise training	Recruiting	12 March 2021
	ISRCTN74393113	Biophysical and psychosocial wellbeing in children with congenital heart disease	Motivational and exercise programme	Completed	29 March 2018
	ISRCTN29534081	Effects of a school-based intervention programme on growth, health, and well-being of schoolchildren in three African countries: the KaziAfya project	School-based health promotion programme	Ongoing	28 April 2021
National Institute of Health, USA	NCT02542683	Physical activity and cognitive development in children	Structured physical activity programme	Recruiting	14 July 2020
	NCT03435354	Enhanced physical activity support in congenital heart disease clinical care (PAToolKit)	Physical activity counselling	Enrolling by invitation	17 July 2020
	NCT04619745	A feasibility study of physical activity after surgical or catheterization intervention	Individualised home and play-based physical activity plans	Recruiting	26 January 2021
	NCT04056416	Physical activity promotion in children and adolescents with single ventricle physiology (MedBike)	MedBike	Enrolling by invitation	4 November 2020
	NCT04106154	Impacting children's physical and mental health through kinesiology support in clinical care	Kinesiology support	Recruiting	17 July 2020
	NCT04208893	Exercise training strategies for children with repaired tetralogy of Fallot	Exercise training	Suspended due to COVID-19 restriction	26 February 2021
	NCT03488797	Web-based motor intervention to increase health related physical fitness in children with congenital heart disease	Supervised web-based and home-based exercise intervention	Completed	5 February 2021
	NCT04815577	Evolution of cardiopulmonary fitness in children with congenital heart disease (Follow-Heart)	Retrospectively compared CPET from 2010 to 2020	Completed	25 March 2021
	NCT04575883	HIIT in youth with congenital heart disease (MedBike)	MedBike	Not yet recruiting	12 April 2021
	NCT04264650	Effectiveness of an mHealth intervention for youth with congenital heart disease	mHealth apps and gamification	Completed	11 February 2020
	NCT02240147	Start-to-sport - home-based exercise for adolescents and adults with congenital heart disease (S2S-ACHD)	Home-based exercise training	Completed	15 January 2019
	NCT01822769	Cardiopulmonary rehabilitation for adolescents and adults with congenital heart disease	Cardiopulmonary rehabilitation	Completed	25 January 2018
	NCT03690518	Rehabilitation of adolescents and young adults with congenital heart diseases (QUALIREHAB)	Cardiac rehabilitation	Recruiting	9 January 2020

CHD, congenital heart disease; CPET, cardiopulmonary exercise test; HIIT, high-intensity interval training; mHealth, mobile health; PA, physical activity.

prescribe PA safely and comprehensively. The small sample sizes in some studies were probably due to restrictive inclusion criteria, such as only limited to specific CHD cohorts and surgical procedures[30 36 37 39 44]; furthermore, only four studies included comparisons with a healthy control group.[35 38 43]

Exercise prescription is only as effective as participants' adherence, which was poorly described in most studies.[67] Therefore, acquiring patient and public involvement prior to any intervention could be beneficial in accommodating patients' and parents' expectations and abilities and increase the chances of higher adherence.[67]

HIIT versus MICT

Traditional exercise programmes involving MICT have been shown to be beneficial in improving PA and exercise capacity, QoL, motor development, and muscular fitness in paediatric patients with CHD[29–47] and adult population.[50–57] However, the relevance of such programmes to the sporadic high-intensity nature of children's play pattern has been questioned.[68 69] HIIT is characterised by short, repeated bouts of vigorous PA (>64%–90% of VO_2 max or 85%–95% peak heart rate (HR)) separated by periods of active recovery at lower intensity.[70 71] Interest in HIIT is topical as it is potentially effective and time-efficient.[68] HIIT reportedly elicits greater training stimulus[72] and subsequently yields equivalent or better increase in exercise capacity than MICT in paediatric healthy[73 74] and obese population (see table 2) and adults with heart failure.[75 76] HIIT has also become an effective exercise intervention for adults with CHD, with several showing greater improvement in exercise capacity following HIIT compared with MICT,[54 77 78] and improvement in flow-mediated vasodilation and pulse wave velocity and reduction in cardiac disease biomarkers N-terminal pro b-type natriuretic peptide (NT-proBNP) and fibrinogen levels.[54] Interest in HIIT continues to grow as it has recently been investigated in children and adolescents with Fontan physiology.[79] Preliminary findings of the study were consistent with those of healthy children,[80] showing that HIIT appears to be safe, feasible and enjoyable.[79] While HIIT has been shown to be safe and well tolerated in children,[62 73 74 79] extensive HIIT studies across various CHD types are warranted as HIIT is being considered an emerging exercise training option that addressed lack of motivation, enjoyment and limited time.[62] The lack of exercise options could be one of the contributing factors towards the underutilisation of exercise training in the paediatric setting.

Home-based versus centre-based exercise intervention

The well-established centre-based exercise intervention (CBEI) is safe and effective in reducing cardiovascular events, hospital readmissions and mortality in patients with CVD.[5 62 81] However, patient participation in CBEI remains low among the elderly, women, children and those of lower socioeconomic status.[81] HBEI was introduced as an alternative, addressing issues such as logistics, transportation and time. It is a form of structured exercise programme with the objective of monitoring and follow-up either through visits, emails, telephone calls, video conferencing or daily diaries of PA.[62 67] A mixture of CBEI and HBEI would be a good transition in shifting the responsibility of the intervention to the patients before HBEI is fully implemented with the aid of advanced telemedicine for early detection of adverse effects.[39 47 50 82] HBEI has been conducted in both adults[77 78 82–86] and children with CHD.[35 36 38 40–46] Unlike adults, children are dependent on parents' availability; thus, minimal travelling makes HBEI preferable for some patients. HBEI in children with CHD has resulted in improvements in maximal oxygen consumption and work rate, muscular strength, PA levels, and health-related QoL, as presented in table 3.

It is difficult to ascertain the overall effectiveness of HBEI and CBEI based on single components, particularly in interventions that involve multiple confounding variables, for example, blood pressure and lipid control, dietary therapy, and psychological support.[81] It is not known whether one or more of these variables have more potential to impact comparably. The diversity of patients' background, duration of a session, exercise intensity and length of the programme may have influenced the results.[81] Conversely, lifestyle and behavioural changes towards PA have been argued to be easier to sustain after the completion of HBEI compared with CBEI due to a higher degree of self-monitoring, but this needs further investigation.[81] One potential advantage of home-based compared with centre-based is that the former offers more flexibility and convenience to patients, which could help to boost participation and long-term adherence.[67] Besides, HBEI may be a timely option for children with CHD to remain physically active during COVID-19 without needless hospital travel.[87]

Monitoring home-based exercise: the use of personal monitoring devices

Commercial activity trackers, also known as 'wearables', are promising tools for measuring, prescribing and promoting PA,[88] with features that include being user-friendly, unobtrusive and affordable. The wearables help to reinforce daily PA goals, sleeping pattern and other behaviours via interactive 'real-time' feedback and reward features,[89] and subsequently increase PA adherence.[10] Recently, the wearables with optical sensors embedded inside of the wrist wrap (photoplethysmography) have been used in clinical paediatric settings and in small cohort study of children with CHD,[10 15 36 88–90] that is, a validity study of wearables against accelerometer,[88] followed by the most recent randomised controlled trial (RCT) comparing PA level between children with CHD and healthy population. The RCT revealed that children with CHD are comparably active with healthy peers ($p=0.217$) and 123 children with CHD (75.9%) have reached 60 min on weekly average of PA according to the WHO criteria.[15] Similarly, 13 of 14 children with Fontan circulation reported that wearables had helped them to monitor PA adherence during HBEI and remain physically active in most days of the week.[36]

Studies in adults revealed that there are discrepancies in the energy expenditure (EE) estimations across different speeds or exercise intensities despite being validated.[89 91] For example, Fitbit One and Vivofit correlated significantly in EE estimation ($p<0.01$; $r=0.702$ and $r=0.854$, respectively) compared with indirect calorimetry (IC) across all gait speeds between walking and running (0.70–3.33 m/s); however, the EE estimation by Vivofit was prone to underestimate, whereas Fitbit One was prone to overestimate.[91] These inconsistencies are probably due to the specific EE equation in the estimation algorithm for each wearable and the placement sites as instructed by the manufacturer.[91] The exaggerated hip movement during walking or running may contribute to higher values when the wearable is placed at the hip compared with at the wrist.[91]

Polar chest strap monitor had the highest agreement in measuring HR compared with ECG ($r_c=0.99$), possibly due to the electrodes embedded in the chest strap to measure cardiac electrical activity.[92] In the same study, Apple Watch was the most accurate wrist-worn wearable in measuring HR, with no statistical difference from ECG ($p=0.22$), whereas the other wearables underestimated the true HR value ($p<0.00001$).[92] The accuracy of HR measurement using wrist-worn wearables over exercise intensities has been trivial and may be reduced at higher intensities.[93 94] Vivoactive HR was reported to have strong correlations ($r=0.96–0.99$, $p<0.05$) in HR measurement during 3 km walking at a steady-state intensity, but only a moderate correlation during interval exercise when compared with Polar chest strap (r=0.58–0.59, p<0.05).[95] This may be the result of variable motion artefacts associated with different exercise modes.[92 93] While some studies reported wearables measuring HR are typically accurate when at rest or walking,[94 96] current evidence shows the accuracy is variable between types of exercise and exercise modalities.[92] Therefore, it is difficult to compare these devices properly, as certain wearables were better suited for certain types of activities or modalities.[92 93 95]

Standardised protocols should be developed further, in particular for HR and EE estimation and under free-living conditions to reflect real-life situation.[97] There is a paucity of research concerning reliability and validity of wearables among children. Disagreements are likely due to different methodologies and the absence of age-appropriate devices. Wearables designed for children used in the two most recent studies[15 98] may add some changes in the assessment of PA among children with and without CHD. Attractive design and interactive gamification features may be crucial factors for long-term compliance during exercise, but this needs to be examined further.[98 99] Although theoretically appealing, the issue remains whether wearables are successful in changing behaviours, and specifically whether they facilitate long-term healthy behavioural change in children with CHD.

CURRENT DIRECTION AND ONGOING CLINICAL TRIALS

As summarised in table 4, 16 ongoing clinical trials have been identified from the National Institute of Health (NIH), USA and International Standard Randomised Controlled Trial Number (ISRCTN) registry using PA and exercise as intervention in paediatric CHD populations. The search criteria used in the registry were 'congenital heart disease' and 'exercise'. The searches were filtered into children and adolescents (<18 years), and only current, recruiting and recently completed studies from 2018 until the most recent update posted in April 2021 were included.

CONCLUSION

Most children born with CHD now live into adulthood, so early prevention, through PA and exercise, is recommended to minimise the risk of long-term complications and comorbidities in later life. There is an accumulating body of evidence which demonstrates that regular PA and exercise are safe and effectively increase PA levels in both healthy and children with CHD. Although there is much uncertainty about the optimal strategy of exercise interventions, addressing the limitations discussed in this review could help to best integrate exercise for the paediatric CHD population. Considering the use of HIIT rather than MICT could be a viable option that is consistent with the nature of children's PA, that is, typically highly intensive but sporadic and appears to be as enjoyable. Home-based intervention is a timely solution to remain physically active at home amid the COVID-19 pandemic, with the added use of wearables to boost long-term PA adherence. Clinicians and paediatricians must strongly advocate the importance of active lifestyle and consider promoting published PA and exercise recommendations, preferably through written individual PA/exercise programmes and aiming for long-term behavioural change. Children who form consistent patterns of healthy PA are therefore more likely to maintain such behaviours as adults.

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