



Physical activity in relation to motor performance, exercise capacity, sports participation, parental perceptions, and overprotection in school aged children with a critical congenital heart defect

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ABSTRACT

Objective: To depict objectively measured moderate-to-vigorous physical activity (MVPA), motor performance (MP), cardiorespiratory fitness (CRF), organized sports participation, parental perceptions of vulnerability and parenting style in children with a Critical Congenital Heart Disease (CCHD), and to explore whether these factors are associated with MVPA.

Study design: A prospective observational cohort study in 62 7–10 years old children with a CCHD.

Results: On average, children with CCHD spent 64 min on MVPA per day (accelerometry), 61 % met the international WHO physical activity guideline. Only 12 % had >60 min of MVPA daily. Eighteen percent had a motor delay (movement-assessment-battery-for children-II) and 38 % showed a below average CRF (cardiopulmonary exercise test using the Godfrey ramp protocol). Seventy-seven percent participated in organized sports activities at least once a week. Twenty-one percent of the parents are classified as overprotective (parent protection scale) and 7.3 % consider their child as being vulnerable (child vulnerability scale). A significant positive association was found between MVPA and MP ($r_s = 0.359$), CRF($VO_{2peak}/ml/kg$: $r_s = 0.472$ and W_{peak}/kg : $r_s = 0.396$) and sports participation ($r_s = 0.286$). Children who were perceived as vulnerable by their parents showed a significantly lower MVPA ($r_s = -0.302$). No significant associations were found between mean MVPA and parental overprotection.

Conclusion: Even though the majority of school aged children with a CCHD is sufficiently active, counseling parents regarding the importance of sufficient MVPA and sports participation, especially in parents who consider their child being vulnerable, could be useful. Since motor delays can be detected at an early age, motor development could be an important target to improve exercise capacity and sports participation to prevent inactivity in children with a CCHD.

Abbreviations: AAA, aortic arch anomaly;; CCHD, critical congenital heart defect; CPET, cardiopulmonary exercise testing; CRF, cardio respiratory fitness; CVS, Child Vulnerability Scale; HRpeak, peak heart rate; Movement-ABC-II, Movement Assessment Battery for Children, 2nd edition; MP, motor performance; MVPA, moderate to vigorous physical activity; PA, physical activity; PPS, Parent Protection Scale; SVP, single ventricle physiology; TGA, transposition of great arteries; ToF, tetralogy of Fallot; VO_{2peak} , maximum oxygen uptake; W_{peak} , maximum workload.

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1. Introduction

Congenital heart disease (CHD) occurs in approximately 9 per 1000 live births, which corresponds to 1.35 million newborns worldwide every year [1]. Around one-fourth of all CHDs who are diagnosed in the antenatal or early neonatal period require cardiac surgery in the first months of life, e.g., transposition of the great arteries (TGA) and tetralogy of Fallot (TOF), hypoplastic left-heart syndrome (HLHS) and are therefore considered critical [2]. Due to improved surgical technology and perioperative management in recent decades, the survival rate of patients with critical congenital heart disease (CCHD) has increased significantly to almost 90 %, leading to a new population at risk for major long-term adverse events [3]. As a result of these improved survival rates, the focus of medical care and research has shifted to improving quality of life and morbidity prevention in patients with a CCHD. Research related to morbidity reduction focuses on neuro-developmental outcomes, cardiorespiratory fitness, and physical activity levels [4–6].

Physical activity (PA) is essential for children's well-being and development, and is considered vital to enhance cardiorespiratory fitness (CRF) and overall health [7]. Higher PA levels have positive, longer-term implications for improving physical and psychosocial health, behavior, and academic performance in children with a CHD [8,9]. The current World Health Organization (WHO) guideline for physical activity recommends children to spend an average of at least 60 min per day of moderate-to-vigorous physical activity (MVPA) across the week [10]. The Dutch PA guideline for children recommends 60 min of MVPA daily [11]. Furthermore, children should incorporate their vigorous-intensity aerobic activities, along with muscle and bone strengthening exercises, for at least 3 days a week to improve or maintain their CRF [10,11]. With the exception of some children with severe postoperative clinical consequences, most children with CCHD do not need exercise restrictions and are advised to comply with these public health recommendations [12].

Despite the important benefits, a significant number of children with CCHD do not meet these PA recommendations [5]. Besides lower activity levels, children with a CCHD, especially those with SVP, often have reduced CRF [13]. Decreased CRF may limit participation in school, sports and social events and could impede future health and quality of life [14]. This is worrisome as both inactivity and reduced exercise capacity are related to survival and health outcome in adults [12].

Some studies mention the role of parental overprotection on physical activity levels of children with a (C)CHD [15]. An overprotective parenting style and parent perceived child vulnerability are considered common in parents who raise a child with a life-threatening or chronic condition [16]. This could impose unnecessary restrictions on children with CCHD due to misconceptions concerning the risks and benefits of physical activity for children with CCHD [15,17,18]. An overprotective parenting style could also contribute to other determinants of PA, like CRF and motor performance (MP) [15,18,19]. In addition, several studies in healthy children indicate a significant association between PA and MP during childhood and through early adolescence, whereby maximum oxygen uptake (VO_{2peak}) mediates this association in both directions [20]. Motor delays are regularly reported in children with a CCHD and are associated with a high prevalence of postoperative brain injury and various other patient-related and procedure-related parameters around neonatal cardiac surgery [21]. The prevalence of children with a motor delay seem to increase when children get older [22].

The number of studies on the incidence and underlying cause of reduced PA in school-aged children (4–12) with CCHD is limited [5,6,23]. Furthermore, no studies are available in school-aged children with a CCHD on the interplay between PA, CRF, MP and the possible role of parental perceptions and parenting style. Given the improved survival and thus increasing number of patients with a CCHD reaching adulthood [3], the need for information on how to oppose inactivity is increasingly

important. Since physical activity habits are already established during childhood [24], understanding the determinants of inactivity in young children with a CCHD could provide useful information for developing early interventions to improve both PA, CRF and MP in the early years.

Therefore, the primary aim of this study is to describe objectively measured moderate-to-vigorous physical activity levels (MVPA), motor performance, cardiorespiratory fitness, organized sports participation, parental perceptions of vulnerability and parenting style in school aged children with CCHD. Secondary, we want to explore whether MVPA is associated with motor performance, cardiorespiratory fitness, organized sports participation, parental perceptions of vulnerability and parenting style in this pediatric population.

2. Materials and methods

2.1. Study sample

This prospective observational cohort study is part of the institutional developmental outpatient follow-up clinic named “*Hart op Weg*” at the Wilhelmina Children's Hospital, University Medical Center Utrecht, Utrecht, the Netherlands. Since July 2011 all children with a CCHD who underwent cardiac surgery with cardiopulmonary bypass within the first six months of life were invited to participate in the developmental follow up as part of standard care. According to the Ethics Committee of the UMC Utrecht this study was not subject to the Medical Research Involving Human Subjects Act (WMO) (reference number 13/442). Signed parental informed consent was obtained for participation in our registry. All children gave verbal consent as well.

2.2. Participants

During the study period (March 2021 – December 2022), 62 patients visited the outpatient clinic for their 7 or 10 years follow up moment. Exclusion criteria for current data analysis included confirmed genetic anomalies such as trisomy 21, 22q11 deletion or CHARGE syndrome as these infants are at increased risk of developmental disorders regardless of their CHD. Because previous studies found significant differences between different diagnosis groups in physical activity [25], motor performance [26], and cardiorespiratory fitness [13], results will be described both for the total group and per diagnostic group. Cardiac diagnoses were grouped as: Transposition of Great Arteries ($n = 32$), Tetralogy of Fallot ($n = 15$), Single Ventricle Physiology ($n = 11$) and Aortic Arch Anomaly ($n = 4$). Significant differences between children with an univentricular repair (SVP) and a biventricular repair (other cardiac diagnoses) will also be described.

2.3. Outcome assessment

2.3.1. Physical activity

MVPA was assessed with the Actigraph GT9X Link accelerometer (ActiGraph Corporation, Pensacola, FL, USA). This waist-worn accelerometer is widely used to objectively measure habitual PA in children [27] and measures accelerations across 3-axis and was worn for 7 consecutive days. Participants were asked to wear the accelerometer on the left hip above the iliac crest, continuously for the next 7 days from 7 a.m. until 9 p.m. It had to be removed only for water-based activities (including showering and swimming) and during sleep. We used ActiLife v.6.13.2 (ActiGraph LLC) for accelerometer initialization (sampling set at 30 Hz) and file download, processing, and analysis. Epoch length was set on 1 s and cut off points by Evenson et al. were used to estimate time spent in sedentary, light, moderate and vigorous intensity activity [27]. If the device was worn ≥ 10 h a day, it was defined as a valid day. Participants' overall accelerometer data were defined as valid if they had ≥ 4 valid days, composed of ≥ 3 weekdays and ≥ 1 weekend days. Compliance with PA guidelines (average ≥ 60 min of MVPA per day) was established by dividing the MVPA (in minutes) of all valid days and

dividing it by the number of valid days. To enable comparison of our results with other studies and healthy peers, our data were compared with both the international WHO PA guideline (average ≥ 60 min of MVPA per day) and the Dutch guideline (daily 60 min MVPA). Besides MVPA in minutes, the mean number of step counts per day and weekly average number of step counts was also reported. The reference goal of 12,000 steps per day was used as this number best corresponds to 60 min of MVPA in children [28].

2.3.2. Motor performance

The movement assessment battery for children – second edition (Movement ABC-II-NL) was used to investigate motor performance. The Movement ABC-II-NL is a global test of motor performance, with assessments of both fine and gross motor coordination. This test is divided into three different age bands. The age band 7–10 was appropriate for all participants of this study. Each age band consists of eight items which are divided into three subsets: manual dexterity, aiming and catching, and balance. Raw scores are converted into standard scores (1–19) and percentile scores (0–100).

Scores above the 16th percentile are regarded as average motor performance. Scores between the 6th and 16th percentile are considered ‘at risk’ for motor difficulties and scores below the 6th percentile indicate significant motor difficulties. Dutch normative data were used to identify children with motor difficulties. The Movement ABC-II-NL has reasonable to good clinical utility in identifying children with motor difficulties [29].

2.3.3. Cardiorespiratory fitness (CRF)

Cardiopulmonary exercise testing (CPET) was used to assess cardiorespiratory fitness (CRF). The patient performed a CPET on an electronically braked cycle ergometer (Lode examiner; Lode BV, Groningen, The Netherlands). Patients breathed through a mouthpiece that was connected to a calibrated metabolic cart (Oxycon Champion; Jaeger, Viasys, Bithoven, The Netherlands). Expired gas was passed through a flow meter, oxygen analyzer, and carbon dioxide analyzer. The flow meter and gas analyzer were connected to a computer, which calculated breath-by-breath minute ventilation, oxygen consumption, carbon dioxide production, and respiratory exchange ratio (RER) from conventional equations. Heart rate (HR), saturation and blood pressure was measured continuously during the maximal exercise test with respectively a 10-lead electrocardiogram. The instrumented participants started with three minutes sitting in rest, followed by a three minutes warming-up of unloaded cycling. Thereafter, the work rate (WR [W]) increased with 10, 15 or 20 W min, dependent on body height according to the Godfrey ramp protocol, until the patient stopped due to volitional exhaustion, despite verbal encouragement. CRF was expressed as peak oxygen uptake (VO_{2peak}) and workload peak (W_{peak}). VO_{2peak} was measured as absolute VO_{2peak} in litres of oxygen per minute and relative VO_{2peak} in milliliters of oxygen per kilogram of body mass per minute ($VO_{2peak/kg}$). VO_{2peak} was measured as the mean value of the last 30 s during the CPET. Relative VO_{2peak} is important for performing activities of daily live and sports since those are weight bearing and helps to control for differences in body mass between subjects. W_{peak} was measured as absolute W_{peak} and relative peak workload per kilogram of body mass per minute ($W_{peak/kg}$). Predicted VO_{2peak} , $VO_{2peak/kg}$, W_{peak} and $W_{peak/kg}$ values were obtained from age- and sex-matched Dutch controls [30]. Further, the ventilatory anaerobic threshold (VAT) is important to sustain endurance activities such as walking or cycling to school. Therefore the VAT was calculated using a computerized v-slope method, using the VCO_2 vs VO_2 plot to detect the beginning of the excess CO_2 output. Z-scores (VO_{2peak} , $VO_{2peak/kg}$, W_{peak} and $W_{peak/kg}$) and percentages of predicted (VAT) values were used to assess the association between PA, CRF and the other outcome variables. The CPET was considered valid if children had a RER >1.0 at peak exercise (RER_{peak}) and/or a peak heartrate (HR_{peak}) of >180 BPM.

2.3.4. Cardiac function

Cardiac function at time of follow up was obtained from patient records. Systolic function of the systematic ventricle was determined by echocardiography and classified as normal, mildly reduced, moderately reduced or severely reduced according to the recommendations of Lopez et al. [31] Cardiac function was only included in the analyses when it was determined within 12 months before or after the Hart op Weg outpatient clinic visits.

2.3.5. Organized sports participation

Information on participation in organized sports activities, (playing on a team or taking lessons within a sport club) as well as the frequency, was verbally obtained.

2.3.6. Parenting style

Parental overprotection is defined as a level of protective parenting behavior that is considered excessive given the child's developmental stage. Parental protective behavior was measured using the parent protection scale (PPS) [32]. The PPS is a 25-item parent self-report scale. Responses were scored using a 4-step scale of 0–3 (0 for “never”, 1 for “sometimes”, 2 for “most of the time”, and 3 for “always”). The total score on the PPS is derived from the sum of all items (possible range 0–75), with higher scores representing greater levels of protection. Norms by child age in the form of cut off points corresponding to the +1 SD (85 %) were used. For 6/7-year-old children the cut-off score is 35 points and for 10 year olds the cut-off score is 29 points. The PPS had acceptable internal consistency, test-retest reliability and clinical validity [32].

Perceived child vulnerability reflects parental attitudes or beliefs that their child is particularly vulnerable or susceptible to harm. Perceived vulnerability of their child was screened with the Child Vulnerability Scale (CVS) [19,33]. The CVS was developed to measure parental perceptions of vulnerability and to identify children perceived as vulnerable. The CVS is an eight-item parent self-report scale. Each of the CVS items is scored on a four-point Likert scale ranging from 0 to 3 (0 = definitely false, 1 = mostly false, 2 = mostly true and 3 = definitely true). Total scores range from 0 to 24, with higher scores reflecting higher perceived vulnerability. A total score equal to or >10 is suggested as a cut-off for high perception of vulnerability in children aged 4 to 8 years. The internal consistency and test-retest reliability of the CVS in children aged 4–8 are adequate [33].

2.4. Statistical analysis

Descriptive statistics (both calculated for the total group and per diagnosis group) were calculated as frequencies, percentages (%), mean and SD, or median (interquartile range [IQR]) depending on the type of data. Normality of distribution of continuous variables was tested with the Kolmogorov–Smirnov test and visualized by skewness and kurtosis of histograms. If not normally distributed, medians and interquartile ranges were reported.

Differences in parametric, non-parametric, and dichotomous outcomes were analyzed using ANOVA and *t*-tests, Kruskal–Wallis or chi-square test. To investigate associations between selected variables spearman's rho, chi-square tests were used. Statistical analyses were performed with IBM SPSS® version 25.0 (SPSS, Chicago, IL). A *p*-value $< .05$ was considered significant.

3. Results

3.1. Patient characteristics

A total of 62 children (43 boys, 19 girls) aged 7 or 10 years old visited our outpatient clinic between February 2021 and December 2022 and fulfilled the inclusion criteria. Reason for exclusion were prematurity ($n = 3$) and an underlying genetic anomaly ($n = 1$). Median age at follow

Table 1
Baseline characteristics of the study cohort.

	Total (N = 62)	TGA (N = 32)	TOF (N = 15)	SVP (N = 11)	AAA (N = 4)
Male n (%)	43 (69.4)	26 (81.3)	10 (66.7)	4 (36.4)	3 (75.0)
Birth weight, grams	3548 ± 595)	3621 ± 530)	3242 ± 776)	3505 (591)	3900 ± 99
Gestational age, weeks	39.4 ± 1.2)	39.6 ± 1.2)	39.4 ± 1.6)	38.9 ± 0.9)	39.3 ± 0.4)
Apgar score 5 min	9 (8–9.75)	8 (8–9)	9 (9–10)	9 (9–10)	9.5 (9–10)
Prenatal diagnosis	43(66.1)	22 (68.8)	7 (46.7)	11 (100)	3 (75.0)
Balloon Atrioseptostomy (BAS) n (%)	21 (33.9)	21 (65.6)	–	–	–
Age at surgery (days)	10 (8–20.25)	9 (7–10)	76 (41.5–89.5)	9.5 (8–22.75)	10 (9–11)
Congenital heart disease, n (%)					
Transposition of Great Arteries (TGA)	32 (51.6)	32 (100.0)	–	–	–
Tetralogy of Fallot (TOF)	15 (16.1)	–	15 (100.0)	–	–
Single Ventricle Physiology (SVP)	11 (17.7)	–	–	11 (100.0)	–
Aortic Arch Anomaly (AAA)	4 (6.5)	–	–	–	4 (100.0)
Primary surgical procedure, n (%)					
Arterial switch	20 (32.3)	26 (81.3)	–	–	–
TOF correction	13 (21.0)	–	13 (86.7)	–	–
Norwood	8 (12.9)	–	–	8 (72.7)	–
AP shunt	6 (9.7)	3(9.4)	1 (6.7)	2(18.2)	–
Aortic arch repair	3 (4.8)	–	–	–	3 (75.0)
Aortic arch repair with aortic valve repair	1 (1.6)	–	–	–	1 (25.0)
VSD closure	1 (1.6)	–	1 (6.7)	–	–
Arterial Switch with VSD closure	7 (11.3)	1 (3.1)	–	–	–
DKS and BT shunt	1 (1.6)	–	–	1 (9.1)	–
Others	2 (3.2)	2 (6.3)	–	–	–
Repeated cardiac surgery n (%)	21 (33.9)	3 (9.4)	4 (26.7)	11 (100)	3 (75.0)
Characteristics at time of assessment					
Systemic Ventricle function n (%)					
- Normal	49 (79.0)	31 (96.9)	10 (66.7)	5 (45.5)	3 (75.0)
- Mildly reduced	11 (17.7)	1 (3.1)	5 (33.3)	4 (36.4)	1 (25.0)
- Moderately reduced	2 (3.2)	–	–	2 (18.2)	–
- Severely reduced	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)
Age at follow-up	7.8 (7.5–7.9)	7.7 (7.5–7.9)	7.9 (7.7–10.2)	7.7 (7.5–7.8)	7.6 (7.1–7.7)
6 years	1 (1.6)	–	–	1 (9.1)	–
7 years	49 (72.0)	28 (87.5)	9 (60.0)	8 (72.7)	4 (100.0)
8 years	3 (4.8)	1 (3.1)	2 (13.3)	–	–
10 years	9 (14.5)	3 (9.4)	4 (26.7)	2 18.2)	–
Height (cm)	128.5 (124.3–135.2)	128.0 (125.2–131.9)	136.5 (126–146.1)	125.0120.6–133.8)	129.6120–139.2)
Weight (kg)	25.7 (23.3–28.9)	25.4 (22.9–27.9)	28.7 (23.4–37.2)	25.2 (23.3–30.4)	25.6 (20–31.2)
BMI (Kg/m ²)	15.9 (14.9–16.5)	15.3(14.9–16.1)	16 (14.5–17.4)	16.2(15.7–16.9)	15 (13.9–16.1)

Data are presented as mean ± standard deviation (* normally distributed) or as median with 25th/75th centiles (not normally distributed) or as number with percentage.

TGA: transposition of great arteries; TOF: tetralogy of Fallot; SVP: single ventricle physiology; AAA: aortic arch anomaly.

up was 7.8 (7.5–7.9). Patient characteristics, including cardiac diagnosis, procedures, and cardiac function at or around time of assessment are presented in Table 1.

3.2. Outcome

3.2.1. Activity levels

Valid accelerometer data were obtained from 49 children (34 boys, 15 girls).

Mean MVPA was 64.1 ± 18.3 min per day. Children with and without valid accelerometer data did not differ significantly in terms of age, diagnosis, gender, cardiac function and sports participation ($p \geq 0.05$). Six children (12.2 %) had >60 min of MVPA on all valid days measured and therefore met the national guideline. Thirty children (61.2 %) met the current international WHO guideline of a weekly average of 60 min MVPA. No significant differences between different diagnostic groups (TGA, TOF, SVP and AAA) or between children with an univentricular repair (SVP) and children with a biventricular repair (TGA, TOF and AAA) were found ($p > .05$).

3.2.2. Motor performance

Sixty-one children completed the motor assessment (43 boys, 18 girls). The mean motor development score, assessed with the Movement ABC-II-NL, was 8.3 ± 2.8 A total motor developmental score ≤ P5 was observed in 11 (18.0 %) children. Ten (16.4 %), 8 (13.1 %), and 9 (14.8 %) children showed a motor delay (≤P5) in the subdomains: manual

dexterity, aiming and catching and balance, respectively. Seventy percent of children who scored below the 5th percentile on manual dexterity, also had a low (< p16) score on 1 or both other domains. For ball skills and balance, this is 50 % and 66.6 % respectively. In children with a manual dexterity score below the 16th percentile, this was in 63.5 % of children combined with a low (<p16) domain score on 1 or both other domains. For ball skills and balance skills this percentage was 46.2 % and 55.4 % respectively.

No significant differences between different diagnostic groups or between children with an univentricular repair and children with a biventricular repair were found ($p > .05$). Detailed information about the distribution of total motor classifications of the total group and per diagnosis group is displayed in Table 2.

3.2.3. Cardiorespiratory fitness

Fifty-four children performed an exercise test (31 boys, 11 girls). Eight children were under control of a pediatric cardiologist at another hospital and no CPET was performed. Twelve out of fifty-four had a RER_{peak} (below 1.0) or HR_{peak} below 180, their data were therefore not considered as maximal. Valid CPET data were therefore obtained in 42 children.

The median W_{peak} was 79.5 Watt (IQR 69.5–101.3). Mean W_{peak/kg} was 3.1 (±0.6) and mean predicted W_{peak/kg} was 101.2 (±19). Seven (16.7 %) of the children scored below 85 % of predicted values.

The maximum heart rate and VAT (both absolute and relative) did not differ significantly between diagnostic groups or between children

Table 2

Activity levels, motor performance, exercise capacity, sports participation and environmental factors.

Outcome measures						p value
Activity levels	Total n = 49	TGA n = 26	TOF n = 12	SVP n = 7	AAA n = 4	
Mean MVPA (minutes/day)	64.1 ± 18.3	69.1 ± 17.9	59.7 ± 17.5	55.5 ± 12.3	60.6 ± 21.1	.23
Daily >60 min MVPA n (%)	6 (12.2)	5 (19.2)	0 (0)	0 (0)	1 (25.0)	
Weekly average > 60 min MVPA n (%)	30 (61.2)	19 (73.1)	6 (50.0)	3 (42.9)	2 (50.0)	
Mean step counts (steps/day)	9927.4 ± 2529.6	10,233.9 ± 2489.3	9503.9 ± 2384.3	9235.2 ± 2676.1	10,416.7 ± 3550.6	.72
Daily >12,000 steps (n)	1 (2.0)	1 (3.8)	0 (0)	0 (0)	0 (0)	
Weekly average > 12,000 steps (n (%))	9 (18.4)	6 (50.0)	1 (8.3)	1 (14.3)	1 (25.0)	
Movement ABC-II-NL scores	Total N = 61	TGA N = 33	TOF N = 14	SVP N = 10	AAA N = 4	
<u>Total motor score</u>	8.3 ± 2.8	8.7 ± 2.6	8.3 ± 2.9	7.1 ± 1.7	8.3 ± 2.8	.41
>P16	37 (60.6)	25 (75.8)	8 (57.1)	2 (20.0)	2 (50.0)	
>P5 ≤ P16	13 (21.3)	4 (12.1)	2 (14.3)	6 (60.0)	1 (25.0)	
≤ P5	11 (18.0)	4 (12.1)	4 (28.6)	2 (20.0)	1 (25.0)	
<u>Domain standard scores</u>						
<u>Manual dexterity</u>	8.7 ± 3.1	9.2 ± 3.4	8.1 ± 2.0	7.5 ± 2.2	9.3 ± 6.3	.38
>P16 n (%)	39 (63.9)	24 (72.7)	8 (57.1)	4 (40.0)	3 (75.0)	
>P5 ≤ P16 n (%)	12 (19.7)	4 (12.1)	4 (28.5)	4 (40.0)	0 (0)	
≤ P5 n (%)	10 (16.4)	5 (15.2)	2 (14.3)	2 (20.0)	1 (25.0)	
<u>Aiming and catching</u>	8.9 ± 5.4	9.2 ± 2.5	8.8 ± 2.0	8.5 ± 1.8	8.0 ± 5.0	.65
>P16 n (%)	48 (78.7)	25 (75.8)	12 (85.7)	8 (80.0)	3 (75.0)	
>P5 ≤ P16 n (%)	5 (8.2)	4 (12.1)	0 (0)	1 (10.0)	0 (0)	
≤ P5 n (%)	8 (13.1)	4 (12.1)	2 (14.3)	1 (10.0)	1 (25.0)	
<u>Balance</u>	8.6 ± 2.8	8.7 ± 2.0	9.1 ± 3.7	7.9 ± 2.3	7.3 ± 5.6	.58
>P16 n (%)	43 (70.4)	25 (75.8)	10 (71.4)	6 (60.0)	2 (50.0)	
>P5 ≤ P16 n (%)	9 (14.8)	5 (15.2)	2 (14.3)	2 (20.0)	1 (25.0)	
≤ P5 n (%)	9 (14.8)	3 (9.1)	2 (14.3)	2 (20.0)	1 (25.0)	
Exercise capacity	Total n = 42	TGA n = 26	TOF n = 8	SVP n = 6	AAA n = 2	
HR _{peak} (BPM)	174.7 ± 13.0	175.9 ± 11.0	179.3 ± 15.4	167.3 ± 15.6	164.5 ± 13.4	.23
W _{peak} (Watt)	79.5 (69.5–101.3)	90 (74.8–102.3)	77 (64.8–118.8)	66.5 (61.0–74.8)	74.5 (64.0–85.0)	.043*
W _{peak} (% pred)	102.3 ± 21.8	110 ± 21	91.4 ± 12.2	83.5 ± 23.6	102.3 ± 21.8	.015*
W _{peak} < 85 % (n)	10 (23.8)	3 (11.5)	3 (37.5)	4 (66.7)	1 (50.0)	
W _{peak/kg} (Watt/kg)	3.1 ± 0.6	3.3 ± 0.5	2.9 ± 0.6	2.5 ± 0.3	3.0 ± 0.6	.007*
W _{peak/kg} (% pred)	101.2 ± 19.0	107.7 ± 17.8	93.8 ± 17.1	83.5 ± 13.4	101.2 ± 19.0	.018*
W _{peak/kg} < 85 % pred (n)	7 (16.7)	2 (7.7)	2 (25.0)	3 (50.0)	0 (0)	
VO _{2peak} (L/min)	1.1 (0.9–1.3)	1.2 (0.9–1.3)	1.1 (0.9–1.5)	0.9 (0.8–0.9)	1.0 (0.9–1.1)	.057
VO _{2peak} (L/min) (% pred)	84.1 ± 15.7	87.8 ± 16.5	83.0 ± 13.8	71.0 ± 9.8	84.1 ± 15.7	.117
VO _{2peak} (L/min <85 % (n)	22 (52.4)	10 (38.5)	3 (37.5)	6 (100)	2 (100)	
VO _{2peak/kg} (ml/kg/min)	40.0 ± 7.7	42.0 ± 7.5	39.5 ± 7.1	31.5 ± 2.7	40.8 ± 9.7	.021*
VO _{2peak/kg} (ml/kg/min) (% pred)	84.1 ± 15.7	94.4 ± 16.7	90.2 ± 12.3	76.3 ± 9.3	91.0 ± 22.8	.095
VO _{2peak/kg} (ml/kg/min) <85 % pred (n)	16 (38.1)	8 (30.8)	3 (37.5)	4 (66.7)	1 (50.0)	
VAT (L/min)	0.59 (0.50–0.69)	0.62 (0.16)	0.58 (0.46–0.65)	0.53 (0.13)	0.58 (0.06)	.662
VAT (L/min) (%VO _{2pred})	50.15 (11.2)	51.3 (10.6)	49 (15.4)	46.8 (8.0)	50.0 (5.7)	.870
VAT <40%VO _{2pred} (n)	7 (16.7)	4 (15.3)	2	1	0 (0)	
Sports participation (n = 62)						.001*
Yes n (%)	48 (77.4)	29 (90.6)	13 (86.7)	5 (45.5)	1 (25.0)	
No n (%)	14 (22.6)	3 (9.4)	2 (13.3)	6 (55.5)	3 (75.0)	
Times a week (median (IQR))	1 (1–2)	2 (1–3)	1 (1–3)	0.5 (0–1)	0 (0–1)	.006*
Environmental factors						
<u>Parent Protection Scale (n = 56)</u>	28 (26–31)	27 (24–31)	28 (27–32)	31 (30–33)	29.5 (27–30.5)	.10
Classified as normal n (%)	44 (78.6)	24 (82.8)	11 (73.3)	5 (55.6)	4 (100)	
Classified as overprotective n (%)	12 (21.4)	5 (17.2)	3 (20.0)	4 (44.4)	0 (0)	
<u>Child Vulnerability Scale (n = 55)</u>	2 (1–5)	2 (0–4)	1.5 (0.8–5.3)	6 (2–9)	2 (2–4)	.12
Vulnerable child (no) n (%)	51 (92.7)	26 (93.8)	13 (93.3)	8 (88.9)	4 (100)	
Vulnerable child (yes) n (%)	4 (7.3)	2 (6.3)	1 (6.7)	1 (11.1)	0 (0)	

Data are presented as mean (±standard deviation) or as number with percentage. Abbreviations: CCHD: critical congenital heart disease; SVP: single ventricle physiology; TGA: transposition of great arteries; AAA: aortic arch anomaly; ToF: tetralogy of Fallot; Movement-ABC-II: Movement Assessment Battery for Children, 2nd edition; HR_{peak}: peak heart rate; W_{peak}: maximum workload; % pred: percentage of predicted; VO_{2peak}: maximum oxygen uptake; MVPA: Moderate to Vigorous Physical Activity; VAT: Ventilator Anaerobic Threshold.

* Statistically significant difference *p*-value < .05.

with an univentricular repair and children with a biventricular repair (*p* > .05).

Median W_{peak} was significant lower in children with an univentricular repair (66.5 (IQR 61.0–75.8)) compared to children with a

biventricular repair (89.5 (IQR 76.6–103.5)) (*p* = .011).

Children with an univentricular repair scored lower than children with a biventricular repair on both W_{peak/kg} (83.5 ± 23.6 vs 105.7 ± 20.8, *p* = .020) and W_{peak/kg} (2.5 ± 0.3 vs 3.2 ± 0.6, *p* = .003) and W_{peak/kg/%}

(83.5 ± 13.3 vs 104.6 ± 18.1, $p = .012$).

Children with an SVP scored lower than children with a TGA on W_{peak} ($p = .007$), $W_{peak\%}$ ($p = .026$), $W_{peak/kg}$ ($p = .005$) and $W_{peak/kg/\%}$ ($p = .019$).

Median VO_{2peak} was 1.1 l/min (0.9–1.3) and mean $VO_{2peak/kg}$ was 40.0 (± 7.7) ml/kg/min. Mean predicted $VO_{2peak/kg}$ was 84.1 (± 15.7) % of predicted. Sixteen (38.1 %) of the children scored below 85 % of predicted values.

Median VO_{2peak} was significant lower in children with an univentricular repair (0.9 (IQR 0.8–0.9)) compared to children with a biventricular repair (1.1 (IQR 0.9–1.3)) ($p = .08$).

Children with an univentricular repair scored lower than children with a biventricular repair on both $VO_{2peak\%}$ (71.0 ± 9.8 vs 86.3 ± 15.5, $p = .025$) and $VO_{2peak/kg}$ (31.5 ± 2.7 vs 41.4 ± 7.3, $p < .001$) and $VO_{2peak/kg/\%}$ (76.3 ± 9.3 vs 93.3 ± 15.7, $p = .014$).

Children with an SVP scored significantly lower than children with a TGA on $VO_{2peak/ml/kg}$ ($p = .011$). Details of the significance levels, absolute values of W_{peak} and VO_{2peak} and the number of children who scored below 85 % of predicted are shown in Table 2.

3.2.4. Organized sports participation

Forty-eight of the 62 children (77.4 %) participated in organized sports activity at least once a week. The median frequency was 1 (1–2). Significant differences were found between diagnosis groups on whether children participated in organized sport activities ($p = .001$) and the frequency children participated in sport activities ($p = .006$). Children with SVP and AAA tend to participate less compared to children with a TGA ($p = .018$ and $p = .035$ respectively) (details in Table 2) and children with an univentricular repair (median 0 (IQR 0–2)) tend to participate less often compared to children with a biventricular repair (median 2 (IQR 1–3)) ($p = .006$).

3.2.5. Parenting style

Fifty-six parents completed the PPS of which 21 % ($n = 12$) of the parents are classified as overprotective. Fifty-five parents completed the child vulnerability scale of which 6.5 % considered their child as being vulnerable ($n = 4$). No differences in parenting style or parental perceptions of child vulnerability in children with different cardiac diagnoses or cardiac function were found. ($p > .05$) Details are displayed in Table 2.

Parents of children with an univentricular repair scored significantly higher than parents of children with a biventricular repair on both the PPS (32.3 ± 6.3 vs 27.8 ± 4.6, $p = .014$) and the CVL (median 3.5 (IQR 1.75–6.75) vs 2 (IQR 0.25–4) $p = .020$).

3.2.6. Cardiac function

Cardiac function was available for all children. Respectively 49, 11, and, 2 children had a normal, mildly reduced, moderately reduced function of the systemic ventricle. None of the children had a severely reduced ventricular function. Predicted W_{peak} was lower in children with mildly reduced cardiac function (86.2 ± 21.2) compared to children with normal cardiac function (106.1 ± 20.5.8) ($p = .018$) Predicted VO_{2peak} was also lower in children with mildly reduced cardiac function (74.4 ± 11.0) compared to children with normal cardiac function (86.4 ± 15.8) ($p = .048$). Lastly, there was a significant difference between cardiac function and whether ($p = .010$) as well as the frequency ($p = .045$) in which children attend an organized sports activity. Of children with normal, mildly reduced or moderately reduced cardiac function at the time of assessment, 84 %, 64 % and 0 % participated in organized sports activities, respectively. No significant differences were found in MVPA, MP and parenting styles between children with different cardiac functions.

3.3. Associations between activity levels, motor performance, cardiorespiratory fitness, sports participation and parental perceptions and parenting style

Average MVPA in minutes per week was significantly associated with motor performance, cardiorespiratory fitness ($VO_{2peak/kg}$ and $W_{peak/kg}$), the number of times children participated in organized sports activities and the child vulnerability score. No significant associations were found between mean MVPA per day and parenting style.

Activity levels measured in mean step counts per day were significantly associated with motor performance, cardiorespiratory fitness ($VO_{2peak/kg}$ and $W_{peak/kg}$) and. No significant associations were found between mean step counts per day and the number of times children participated in organized sports activities, the child vulnerability score and parent protection scale score ($p > .05$). Details on associations between activity levels, motor performance, cardiorespiratory fitness, sports participation, and environmental factors are shown in Table 3.

4. Discussion

Our aim was to describe objectively measured MVPA, motor performance, CRF, organized sports participation, parental perceptions of vulnerability and parenting style in school aged children with CCHD and to explore whether these factors are associated with MVPA.

4.1. Activity levels

Mean MVPA was 64.1 min/day (±18.3) and 61.2 % met the current international guideline [10] of an average 60 min of MVPA. Only 12.2 % of the patients met the Dutch PA guideline that recommends a daily MVPA of 60 min per day [11]. This is lower compared to healthy Dutch peers: where 62.3 % met the Dutch PA guidelines [34]. The difference might be explained by the fact that MVPA was measured with a questionnaire, which is often answered more positively than objectively measured MVPA [25]. Activity levels in our population (mean MVPA of 64.1 min/day) were higher compared to previous studies who reported that children with CCHD were generally less active than healthy peers [5]. Voss et al. found a median daily MVPA of 43 min/day. Eight percent of their population met the (previous) WHO MVPA guidelines of 60 min MVPA daily versus 12.2 % of our patients met the Dutch PA guideline (60 min MVPA daily). The difference could be explained by the mean age of the populations, as a decline in activity levels with age is common. [5]

No significant differences in MVPA in relation to the diagnostic groups were found. This is consistent with a recent systematic review of Skovdahl et al. and Voss et al. [5,35] A reason for this could be that in healthy children, where determinants of PA were widely studied, the explanation for inactivity appears to be multifactorial [36]. Ecological models posit that besides individual (psychological and biological) factors also interpersonal (social support, cultural norms) and environmental (access to opportunities and facilities), regional or national and even global policy are important determinants of physical activity and are thought to have widespread effects [36]. Our study only investigated a limited number of these factors. A combination of the above factors on PA levels in children with CCHD may have a greater effect than their heart defect itself.

4.2. Motor performance and MVPA

A significant proportion of the children scored at risk ($>P5 \leq P16$) or even delayed ($\leq P5$) on the Movement ABC-II-NL. Eighteen percent scored $\leq P5$. This motor delay is 3 times as high compared to healthy peers. On the total motor scores, manual dexterity, aiming and catching, and balance skills, 21–39 % scored below the 16th percentile, which is normally expected in 16 % of the population.

The majority (70 % and 66.7 % respectively) of the children with a

Table 3

Associations between activity levels, motor performance, exercise capacity, sports participation and environmental factors.

	MVPA min/ wk n = 49	MVPA steps/ wk n = 49	M-ABC total n = 61	Wpeak n = 42	Wpeak/ kg n = 42	VAT% VO ₂ pred. n = 42	VO ₂ peak l/min n = 42	VO ₂ peak ml/min/ kg n = 42	Times a week n = 62	PPS score n = 56	CVS score n = 55
Average MVPA min (n = 49)	–										
Average steps/week (n = 49)	0.874**	–									
M-ABC-II Total (n = 61)	0.359*	0.329*	–								
Wpeak (n = 42)	0.942	0.414	0.350	–							
Wpeak/kg (n = 42)	0.396*	0.362*	0.656**	0.474**	0.396*	0.033					
VAT%VO ₂ pred (n = 40)	0.019	0.033	0.000	0.002	0.019	0.844					
VO ₂ peak (n = 42)	–0.34	–0.093	–0.092	0.224	–0.220	–					
VO ₂ peak/kg (n = 42)	0.854	0.613	0.578	0.225	0.178						
Sports participation/times a week (n = 62)	0.182	0.166	0.185	0.675**	0.406*	0.464**	–				
Parent protection scale (PPS) score (n = 56)	0.303	0.348	0.296	0.000	0.017	0.009					
Child Vulnerability (CVS) scale score (n = 55)	0.472**	0.527**	0.491*	0.231	0.732**	0.082	0.521**	–			
	0.004	0.001	0.001	0.140	0.000	0.621	0.000				
	0.286*	0.274	0.315*	0.234	0.361*	0.144	0.280	0.308*	–		
	0.046	0.057	0.014	0.184	0.019	0.380	0.109	0.047			
	–0.243	–0.086	–0.212	–0.200	–0.318*	–0.036	–0.018	–0.105	–0.048	–	
	0.104	0.568	0.120	0.258	0.049	0.836	0.922	0.523	0.723		
	–0.302*	–0.234	–0.229	–0.194	–0.190	–0.346	–0.084	–0.231	–0.359**	0.309*	–
	0.043	0.121	0.096	0.272	0.253	0.042	0.635	0.163	0.007	0.022	

Numbers in italic represent partial correlations corrected for age. Bold numbers represent significant linear association indicated by Spearman's Rho ($p < .05$). Abbreviations: MVPA: Moderate to Vigorous Physical Activity M-ABC-II-Total: Movement Assessment Battery for Children—second edition total scale score; W_{peak}: maximum workload; W_{peak}/kg: Wpeak corrected for weight; VAT%VO₂pred: maximum oxygen uptake on anaerobic threshold as percentage of predicted. VO₂peak: maximum oxygen uptake; VO₂peak: VO₂peak in liters per minute; VO₂peak/kg: VO₂peak in milliliters per minute corrected for weight; PPS: parent protection scale; CVS: child vulnerability scale.

* Correlation is significant at the 0.05 level (2-tailed).

** Correlation is significant at the 0.01 level (2-tailed).

low (<P5) score on manual dexterity or ball skills, also achieved a low score (<P16) on 1 or even 2 of the other domains. The majority of children who scored low on manual dexterity or ball skills therefore had more generalized motor deficits. Children with low scores (<P5) on ball skills most often had an isolated deficit (50 %).

An explanation for the high number of children with a generalized motor delay might be acquired brain injury around neonatal surgery [37]. Several studies in infants and toddlers have reported a high prevalence of postoperative brain injury and its association with various parameters, which can affect motor development [21,37–40]. The large number of children with lower motor scores (especially on fine motor skills) could also be explained by deficits in executive functions, as both are related and frequently reported in children and adolescents with CCHD, and in particular children with SVP. For example, the fine motor skills tasks of the Movement ABC- II include both precision tasks and speed tasks. In addition to motor coordination, these tasks also require sufficient task orientation, planning, inhibition, attention and concentration which all belong to the “higher cognitive functions” or executive functions [41–45].

In contrast to previous studies within the same population but at a younger age, [22,46] the current study found no significant differences in motor functioning between diagnosis groups. The effect of the type of heart defect or univentricular of biventricular repair on motor performance therefore seems limited (at this age) and possibly other factors play a more important role.

MVPA was found to be significantly associated with motor performance. This association has been described previously in healthy children, but not yet in children with a CCHD [20,47]. This may be explained by the fact that children with better motor skills are more often involved in more physical activities and sports, while the lower motor skills of children increasingly withdraw from these types of activities due to, for example, a lower self-efficacy about their physical performance. [48] Our finding that higher motor performance was

associated with a higher frequency of participation in organized sports activities partially confirms this hypothesis. Children who exercise more often have a better motor performance, but also vice versa. Exercise capacity could influence this association because strong significant associations were also found between sport frequency and exercise capacity as well as motor performance and exercise capacity. This is in line with previous studies in healthy children which indicate a significant association between physical activity and motor performance (MP) during childhood to early adolescence whereby VO₂peak mediates this association in both directions. [20,47]

4.3. Exercise capacity and MVPA

In our study VO₂peak as percentage of predicted was 84 % of predicted for both VO₂peak and VO₂peak/kg. VO₂peak/kg was <85 % of predicted in 38 % of the population. This is in line with a recent study of van Genuchten et al. who found a lower CRF as shown by reduced VO₂peak and W_{peak} in a Dutch sample of children with a CHD compared to healthy Dutch peers [13]. The absolute and relative W_{peak} (corrected for bodyweight) were 102 and 101 % of predicted, respectively. The majority (83 %) of the children achieved a W_{peak}/kg \geq 85 % of predicted. This is not consistent with the results of van Genuchten et al. [13] and a systematic review of Villaseca-Rojas et al. [49] who found lower exercise capacity as shown by reduced VO₂peak, W_{peak}, VE/VCO₂ slope, O₂ pulse, and HRmax in children with a CHD compared with matched healthy controls. There is no obvious explanation for the difference in findings. The participants in our study are, as part of standard care, followed since birth at the neurodevelopmental outpatient clinic. PA counseling is part of all patient interactions in order to make parents aware of the PA recommendations and to inform parents and patients about the importance of sufficient PA. This encouraging approach might explain this different finding in part.

MVPA was found to be significantly associated with CRF for both

$VO_{2peak/kg}$ and $W_{peak/kg}$. This is in line with Ortega et al. who found activity to be associated with healthier CRF levels in adolescents [50,51]. Furthermore, significant differences in CRF between diagnostic groups of CCHD were found. CRF was significantly lower in children with an SVP compared to children with a TGA on both W_{peak} and VO_{2peak} . Of the children with a SVP, 67 % had a $VO_{2peak/kg} < 85$ % and 50 % had a $W_{peak/kg} < 85$ % or predicted. These low values can be explained by the abnormal physiology of the heart of children with SVP after the Fontan operation. As a result, these patients are more limited in their ability to adjust their pulmonary blood flow and blood pressure during exercise which can lead to insufficient ventricular preload and systemic blood flow. In addition, as a result of shunting in response to exercise, children with an SVP often also have a lower maximum heart rate and a lower arterial oxygen saturation.

4.4. Sports participation and MVPA

MVPA was found to be significantly associated with sports participation, expressed in times per week. In our population 77 % of all participants took part in sports at least once a week. Arvidsson et al. [52] reported higher percentages (80–94 %), however, this population also included older children with less severe heart defects. Another explanation for the difference might be that our data were partly collected during the corona pandemic. Restrictions on mobility, social distancing, or closure of schools, and recreational centers, although an effective measure against the spread of COVID-19, might have influenced sports participation and activity levels of the population. In retrospect, it is difficult to determine to what extent COVID-19 has influenced these figures. We found that children with an SVP and AAA participate less often in sports activities compared to children with a TGA. Establishing determinants of sports participation was outside the scope of this study. Since significant differences were found on the PPS and the CVL between children with an SVP and the other diagnostic groups, and significant correlations were also found between the CVL and PPS as well as the CFS and sports participation in this study, the reduced sports participation could possibly be explained by the relatively high scores on the PPS and the CVL in children with an SVP. These relationships were not investigated within this study, but could be an interesting topic for future research within a larger population.

4.5. Parental perceptions and parental overprotection and MVPA

Regarding the role of parents, 21 % of parents showed an over-protective parenting style.

According to the reference values, an increased parental protection score is normally expected in 15 % of the population [32]. In a recent study [53] in children with cancer, 15.6 % scored in the subclinical area of the parent protection scale, so the percentage of parents with an overprotective parenting style seems slightly increased in our population compared to other chronic conditions.

However, no direct association was found between parental overprotection and activity levels. This contrasts with the frequently cited model by Bjarnasson-Wehrens, who described possible interactions between motor development, parental overprotection, and physical activity levels [15].

Lastly, in this population 7 % of parents scored above the cutoff of 10 points on the CVS and perceive their child with CCHD as vulnerable. Forsyth et al. report that 3 % of children (age 4–8) without any medical condition have scores above this cut-off compared to 41 % of the children with a medical condition [33]. Although the number of parents perceiving their child as vulnerable is low, children of parents who see their child as vulnerable participated in sports activities less often and were consequently less physically active (expressed in average number of minutes per week).

4.6. Strengths & limitations

To our knowledge, this is the first study that objectively describes MVPA levels and the role of motor performance, cardiorespiratory fitness, organized sports participation, parental perceptions of vulnerability and parenting style on MVPA in school aged children with a CCHD.

Our study investigated a limited number of individual and interpersonal factors and found only weak to moderate associations between MVPA, motor performance, CRF, parental perceptions of child vulnerability, and sports participation. This suggests, as mentioned before, that other determinants also contribute to children's MVPA [36,52,54]. Finally, to get an idea of whether the different diagnostic groups differ from each other, we described the results not only for the total group, but also for specific types of CCHD. These results should be interpreted with some caution because they concern small groups. A (multi-center) study with a larger patient population will be needed to overcome these "limitations" and could provide important information to influence PA in an even more targeted way.

4.7. Recommendations

Besides the description of the results of all included children, we additionally reported the results per diagnostic group, as in our previous studies significant differences were found between the different diagnostic groups. [22,37,46,55] Analyzing determinants of PA per specific diagnostic group was beyond the scope of the current study, but might be interesting in future studies with larger diagnostic subgroups. Since the TOF population seem to differ from others on several factors (e.g., age at surgery, likelihood of prenatal diagnosis, birth weight, etc.) and children with an SVP score lower on several domains than the other diagnostic groups, it may be considered to classify the patient groups differently, for example univentricular vs biventricular repair or TOF vs the other CCHD's in future studies.

Within this study, children with an underlying genetic anomaly and preterm children were excluded from the data analysis. A future study on the long-term outcomes of children with a CCHD, where differences between full-term and preterm children and children with and without genetic anomalies are investigated, would be interesting as these factors, in addition to the heart defect itself, might significantly influence the long-term outcomes.

Although, several studies on intervention programs to improve CRF and activity levels in children and adults are available [56,57], effective interventions to improve both cardiorespiratory fitness and activity levels in children with a CCHD are scarce [58]. With the exception of a study by Longmuir et al. in children with an SVP [59], motor performance is usually not the focus of interventions to improve CRF or PA. Based on the current study, motor performance may be an important target for early interventions in children with a CCHD to improve both PA and CRF.

An interesting concept that could be used to better understand the individual and interpersonal determinants of PA in children with CCHD is the concept of 'physical literacy' [47]. Higher levels of physical literacy in healthy children are associated with higher levels of physical activity. Besides physical competence (motor performance and cardiorespiratory fitness), self-efficacy, motivation, confidence, understanding, and knowledge are essential to remain physically active [60]. and should be considered in future studies on (improving) activity levels in children with a CCHD as well [61].

While much emphasis is placed on lifestyle medicine in adult cardiology, it is still not often a focus in the pediatric cardiology outpatient clinic [62]. In a previous study pediatric cardiologists indicated that they do not have sufficient knowledge, skills, and time to delve into this sufficiently and to pay attention to it during the outpatient clinics [63]. With regard to knowledge and parental perceptions, clinicians should educate patients and parents about the importance and benefits of

adequate PA, and especially the risks of inactivity. They should be actively made aware of the PA recommendations, and even if there are no PA restrictions, this should also be explicitly mentioned to prevent parents from unnecessarily considering their child as vulnerable.

Given the known risks associated with CCHD and the demonstrated benefits of early intervention in other populations, regular monitoring and neurodevelopmental evaluation up to 60 months is strongly recommended to optimize the neurodevelopmental outcomes and quality of life of patients with CCHD [64]. To protect their current and future cardiovascular health, structural monitoring of PA, CRF and motor development from school age and beyond would be a useful addition to the ongoing health evaluation of children with CHD.

4.8. Conclusion

The majority of children with CCHD are sufficiently active and show age-appropriate motor performance, cardiorespiratory fitness and sports participation. The minority of parents consider their child to be vulnerable or have an overprotective parenting style.

A significant association was found between MVPA and motor performance, exercise capacity, sports participation, and parental perceptions of vulnerability. The role of an overprotective parenting style could not be confirmed in this study. Since motor deficits can be detected at an early age, motor performance could be an important target for intervention to improve exercise capacity and sports participation in order to prevent inactivity in children with a CCHD. Counseling parents regarding the importance of physical activity, especially in parents who consider their child to be vulnerable, could be useful.

Declaration of competing interest

The authors have no conflict of interest to disclose.

Data availability

Data are available from the corresponding author: The authors would like to thank the patients and their parents who participated in the study and all those involved in the Hart op Weg outpatient clinic and the Congenital Heart Disease Life Span research group. The study was made possible through financial support from the Hartekind foundation and the Stichting Vrienden van het Wilhelmina Kinderziekenhuis. The funding sources were not involved in the conduct of the research or preparation of the article.

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Financial disclosure

The authors have no financial relationships relevant to this article to disclose.

Institutional review board statement

This study was not subject to the Medical Research Involving Human

Subjects Act (WMO) according to the Ethics Committee of University of Utrecht (reference number 13/442; 5 September 2013).

Informed consent statement

Informed consent was obtained from all subjects involved in the study. Signed parental informed consent was obtained for participation in our registry.

References

- [1] Y. Liu, S. Chen, L. Zu, et al., Global birth prevalence of congenital heart defects 1970–2017: updated systematic review and meta-analysis of 260 studies, *Int. J. Epidemiol.* 0 (0) (2019) 1–9, <https://doi.org/10.1093/zoolinnea/zly093>.
- [2] W.T. Mahle, J.W. Newburger, G.P. Matherne, et al., Role of pulse oximetry in examining newborns for congenital heart disease: a scientific statement from the AHA and AAP, *Pediatrics* (2009), <https://doi.org/10.1542/peds.2009-1397>. Published online.
- [3] A.J. Marelli, R. Ionescu-Ittu, A.S. Mackie, L. Guo, N. Dendukuri, M. Kaouache, Lifetime prevalence of congenital heart disease in the general population from 2000 to 2010, *Circulation* (2014), <https://doi.org/10.1161/CIRCULATIONAHA.113.008396>. Published online.
- [4] B. Latal, Neurodevelopmental outcomes of the child with congenital heart disease, *Clin. Perinatol.* 43 (1) (2016) 173–185, <https://doi.org/10.1016/j.clp.2015.11.012>.
- [5] C. Voss, S.L. Duncombe, P.H. Dean, A.M. de Souza, K.C. Harris, Physical activity and sedentary behavior in children with congenital heart disease, *J. Am. Heart Assoc.* 6 (3) (2017), <https://doi.org/10.1161/JAHA.116.004665>.
- [6] L. Banks, S. Rosenthal, C. Manhiot, et al., Exercise capacity and self-efficacy are associated with moderate-to-vigorous intensity physical activity in children with congenital heart disease, *Pediatr. Cardiol.* 38 (6) (2017) 1206–1214, <https://doi.org/10.1007/s00246-017-1645-2>.
- [7] I. Janssen, A.G. LeBlanc, Systematic review of the health benefits of physical activity and fitness in school-aged children and youth, *Int. J. Behav. Nutr. Phys. Act.* 7 (2010), <https://doi.org/10.1186/1479-5868-7-40>.
- [8] K. Dulfer, W.A. Helbing, N. Duppen, E.M.W.J. Utens, Associations between exercise capacity, physical activity, and psychosocial functioning in children with congenital heart disease: a systematic review, *Eur. J. Prev. Cardiol.* (2014), <https://doi.org/10.1177/2047487313494030>. Published online.
- [9] D.E.R. Warburton, S.S.D. Bredin, Health benefits of physical activity: a systematic review of current systematic reviews, *Curr. Opin. Cardiol.* (2017), <https://doi.org/10.1097/HCO.0000000000000437>. Published online.
- [10] A.D. Okely, A. Kontsevaya, J. Ng, C. Abdeta, 2020 WHO guidelines on physical activity and sedentary behavior, in: *Sports Medicine and Health Science*, 2021, <https://doi.org/10.1016/j.smhs.2021.05.001>. Published online.
- [11] R.M. Weggemans, F.J.G. Backx, L. Borghouts, et al., The 2017 Dutch physical activity guidelines, *Int. J. Behav. Nutr. Phys. Act.* (2018), <https://doi.org/10.1186/s12966-018-0661-9>. Published online.
- [12] T. Takken, A. Giardini, T. Reybrouck, et al., Recommendations for physical activity, recreation sport, and exercise training in paediatric patients with congenital heart disease: a report from the Exercise, Basic & Translational Research Section of the European Association of Cardiovascular Prevention and Rehabilitation, the European Congenital Heart and Lung Exercise Group, and the Association for European Paediatric Cardiology, *Eur. J. Prev. Cardiol.* 19 (5) (2012), <https://doi.org/10.1177/1741826711420000>.
- [13] W.J. van Genuchten, W.A. Helbing, A.D.J. Ten Harkel, et al., Exercise capacity in a cohort of children with congenital heart disease, *Eur. J. Pediatr.* 182 (1) (2023) 295–306, <https://doi.org/10.1007/s00431-022-04648-9>.
- [14] R. Acosta-Dighero, R. Torres-Castro, I. Rodríguez-Núñez, et al., Physical activity assessments in children with congenital heart disease: a systematic review, *Acta Paediatr.* (2020), <https://doi.org/10.1111/apa.15478>. Published online.
- [15] B. Bjarnason-Wehrens, S. Dordel, S. Schickendantz, et al., Motor development in children with congenital cardiac diseases compared to their healthy peers, *Cardiol. Young* 17 (5) (2007) 487–498, <https://doi.org/10.1017/S1047951107001023>.
- [16] B. Bjarnason-Wehrens, S. Schmitz, S. Dordel, Motor development in children with congenital cardiac diseases, *Eur. Cardiol. Rev.* 4 (2) (2008) 92, <https://doi.org/10.1542/ecr.2008.4.2.92>.
- [17] K. Uzark, K. Jones, J. Slusher, C.A. Limbers, T.M. Burwinkle, J.W. Varni, Quality of life in children with heart disease as perceived by children and parents, *Pediatrics* (2008), <https://doi.org/10.1542/peds.2006-3778>. Published online.
- [18] P.E. Longmuir, B.W. McCrindle, et al., *Am. Heart J.* (2009), <https://doi.org/10.1016/j.ahj.2009.02.014>. Published online.
- [19] B.A. Houtzager, E.L. Möller, H. Maurice-Stam, B.F. Last, M.A. Grootenhuys, Parental perceptions of child vulnerability in a community-based sample: association with chronic illness and health-related quality of life, *J. Child Health Care* (2015), <https://doi.org/10.1177/1367493514530954>. Published online.
- [20] S. King-Dowling, N.A. Proudfoot, J. Cairney, B.W. Timmons, Motor competence, physical activity, and fitness across early childhood, *Med. Sci. Sports Exerc.* (2020) 1–30, <https://doi.org/10.1249/MSS.0000000000002388>. Published online May.

- [21] H.H. Hövels-Gürich, Factors influencing neurodevelopment after cardiac surgery during infancy, *Front. Pediatr.* 4 (December) (2016) 1–6, <https://doi.org/10.3389/fped.2016.00137>.
- [22] M.C.A. Sprong, B.C.H. Huijgen, L.S. de Vries, et al., Early determinants of adverse motor outcomes in preschool children with a critical congenital heart defect, *J. Clin. Med.* (2022), <https://doi.org/10.3390/jcm11185464>. Published online.
- [23] E. Barbour-Tuck, N.G. Boyes, C.R. Tomczak, et al., A cardiovascular disease risk factor in children with congenital heart disease: Unmasking elevated waist circumference—a CHAMPS*study*CHAMPS: Children's Healthy-Heart Activity Monitoring Program in Saskatchewan, *BMC Cardiovasc. Disord.* (2020), <https://doi.org/10.1186/s12872-020-01508-y>. Published online.
- [24] R. Telama, Tracking of physical activity from childhood to adulthood: a review, *Obes. Facts* (2009), <https://doi.org/10.1159/000222244>. Published online.
- [25] L. Brudy, J. Hock, A.L. Häcker, et al., Children with congenital heart disease are active but need to keep moving: a cross-sectional study using wrist-worn physical activity trackers, *J. Pediatr.* (2020), <https://doi.org/10.1016/j.jpeds.2019.09.077>. Published online.
- [26] M.C.A. Sprong, W. Broeders, J. van der Net, et al., Motor developmental delay after cardiac surgery in children with a critical congenital heart defect: a systematic literature review and meta-analysis, *pediatric ph* 33 (4) (2021) 186–197. https://journals.lww.com/pedpt/Fulltext/2021/10000/Motor_Developmental_Delay_After_Cardiac_Surgery.in.3.aspx.
- [27] S.G. Trost, P.D. Loprinzi, R. Moore, K.A. Pfeiffer, Comparison of accelerometer cut points for predicting activity intensity in youth, *Med. Sci. Sports Exerc.* 43 (7) (2011), <https://doi.org/10.1249/MSS.0b013e318206476e>.
- [28] R.C. Colley, I. Janssen, M.S. Tremblay, Daily step target to measure adherence to physical activity guidelines in children, *Med. Sci. Sports Exerc.* (2012), <https://doi.org/10.1249/MSS.0b013e31823f23b1>. Published online.
- [29] B.C.M. Smits-Engelsman, A.S. Niemeijer, H. van Waelvelde, Is the movement assessment battery for children-2nd edition a reliable instrument to measure motor performance in 3 year old children? *Res. Dev. Disabil.* (2011) <https://doi.org/10.1016/j.ridd.2011.01.031>. Published online.
- [30] T. Takken, B.C. Bongers, M. Van Brussel, E.A. Haapala, E.H.J. Hulzebos, Cardiopulmonary exercise testing in pediatrics, in: *Annals of the American Thoracic Society*, 2017, <https://doi.org/10.1513/AnnalsATS.201611-912FR>.
- [31] L. Lopez, S.D. Colan, P.C. Frommelt, et al., Recommendations for quantification methods during the performance of a pediatric echocardiogram: a report from the Pediatric Measurements Writing Group of the American Society of Echocardiography Pediatric and Congenital Heart Disease Council, *J. Am. Soc. Echocardiogr.* 23 (5) (2010), <https://doi.org/10.1016/j.echo.2010.03.019>.
- [32] M. Thomasgard, W.P. Metz, C. Edelbrock, J.P. Shonkoff, Parent-child relationship disorders. Part I. Parental overprotection and the development of the Parent Protection Scale, *J. Dev. Behav. Pediatr.* (1995), <https://doi.org/10.1097/00004703-199508000-00006>. Published online.
- [33] B.W.C. Forsyth, S.M.C. Horwitz, J.M. Leventhal, J. Burger, P.J. Leaf, The child vulnerability scale: an instrument to measure parental perceptions of child vulnerability, *J. Pediatr. Psychol.* (1996), <https://doi.org/10.1093/jpepsy/21.1.89>. Published online.
- [34] Nederlands Jeugd Instituut, Cijfers over beweging, Published, <https://www.nji.nl/cijfers/beweging>, 2022.
- [35] P. Skovdahl, C. Kjellberg Olofsson, D. Arvidsson, Physical activity in children and adolescents with CHD: Review from a measurement methodological perspective, *Cardiol. Young* 31 (4) (2021), <https://doi.org/10.1017/S1047951121000627>.
- [36] A.E. Bauman, R.S. Reis, J.F. Sallis, et al., Correlates of physical activity: why are some people physically active and others not? *Lancet* (2012) [https://doi.org/10.1016/S0140-6736\(12\)60735-1](https://doi.org/10.1016/S0140-6736(12)60735-1). Published online.
- [37] R. Stegeman, M.C.A. Sprong, J.M.P.J. Breur, et al., Early motor outcomes in infants with critical congenital heart disease are related to neonatal brain development and brain injury, *Dev. Med. Child Neurol.* (2021), <https://doi.org/10.1111/dmnc.15024>. Published online.
- [38] S. Peyvandi, B. Latal, S.P. Miller, P.S. McQuillen, The neonatal brain in critical congenital heart disease: insights and future directions, *Neuroimage*. 185 (2019) 776–782, <https://doi.org/10.1016/j.neuroimage.2018.05.045>.
- [39] N.H.P. Claessens, J.M.P.J. Breur, F. Groenendaal, et al., Brain microstructural development in neonates with critical congenital heart disease: an atlas-based diffusion tensor imaging study, *Neuroimage Clin.* 21 (2019) 101672, <https://doi.org/10.1016/j.nicl.2019.101672>.
- [40] P.S. McQuillen, A.J. Barkovich, S.E.G. Hamrick, et al., Temporal and anatomic risk profile of brain injury with neonatal repair of congenital heart defects, *Stroke* 38 (2 PART 2) (2007) 736–741, <https://doi.org/10.1161/01.STR.0000247941.41234.90>.
- [41] I.M.J. van der Fels, J. Smith, A.G.M. de Bruijn, et al., Relations between gross motor skills and executive functions, controlling for the role of information processing and lapses of attention in 8–10 year old children. *Capio CM*, ed, *PLoS One* 14 (10) (2019), e0224219, <https://doi.org/10.1371/journal.pone.0224219>.
- [42] C. Sterken, J. Lemièr, G. Van Den Bergh, D. Mesotten, Neurocognitive development after pediatric heart surgery, *Pediatrics* 137 (6) (2016), <https://doi.org/10.1542/peds.2015-4675>.
- [43] J.H. Sanz, M.M. Berl, A.C. Armour, J. Wang, Y.I. Cheng, M.T. Donofrio, Prevalence and pattern of executive dysfunction in school age children with congenital heart disease, *Congenit. Heart Dis.* (2017), <https://doi.org/10.1111/chd.12427>. Published online.
- [44] A. Diamond, Executive functions, in: *Handbook of Clinical Neurology*, 2020, <https://doi.org/10.1016/B978-0-444-64150-2.00020-4>.
- [45] C.L. Brosig, L. Bear, S. Allen, et al., Neurodevelopmental outcomes at 2 and 4 years in children with congenital heart disease, *Congenit. Heart Dis.* 13 (5) (2018) 700–705, <https://doi.org/10.1111/chd.12632>.
- [46] M.C.A. Sprong, M. van Brussel, L.S. de Vries, et al., Longitudinal motor-developmental outcomes in infants with a critical congenital heart defect, *Children*. 9 (4) (2022) 570, <https://doi.org/10.3390/children9040570>.
- [47] R.A. Lima, K. Pfeiffer, L.R. Larsen, et al., Physical activity and motor competence present a positive reciprocal longitudinal relationship across childhood and early adolescence, *J. Phys. Act. Health* (2017), <https://doi.org/10.1123/jpah.2016-0473>. Published online.
- [48] J. Cairney, J.A. Hay, B.E. Faught, T.J. Wade, L. Corna, A. Flouris, Developmental coordination disorder, generalized self-efficacy toward physical activity, and participation in organized and free play activities, *J. Pediatr.* (2005), <https://doi.org/10.1016/j.jpeds.2005.05.013>. Published online.
- [49] Y. Villaseca-Rojas, J. Varela-Melo, R. Torres-Castro, et al., Exercise capacity in children and adolescents with congenital heart disease: a systematic review and meta-analysis, *Front. Cardiovasc. Med.* (2022), <https://doi.org/10.3389/fcvm.2022.874700>. Published online.
- [50] F.B. Ortega, J.R. Ruiz, M.J. Castillo, M. Sjöström, Physical fitness in childhood and adolescence: a powerful marker of health, *Int. J. Obes.* 32 (1) (2008), <https://doi.org/10.1038/sj.jco.0803774>.
- [51] F.B. Ortega, J.R. Ruiz, A. Hurtig-Wennlöf, M. Sjöström, Physically active adolescents are more likely to have a healthier cardiovascular fitness level independently of their adiposity status. *The European Youth Heart Study*, *Rev. Esp. Cardiol. (Engl. Ed.)* 61 (2) (2008), [https://doi.org/10.1016/s1885-5857\(08\)60087-0](https://doi.org/10.1016/s1885-5857(08)60087-0).
- [52] D. Arvidsson, F. Slinde, L. Hulthén, J. Sunnegårdh, Physical activity, sports participation and aerobic fitness in children who have undergone surgery for congenital heart defects, *Acta Paediatr.* 98 (9) (2009) 1475–1482, <https://doi.org/10.1111/j.1651-2227.2009.01369.x>.
- [53] C.J.M. Colletti, C. Wolfe-Christensen, M.Y. Carpentier, et al., The relationship of parental overprotection, perceived vulnerability, and parenting stress to behavioral, emotional, and social adjustment in children with cancer, *Pediatr. Blood Cancer* (2008), <https://doi.org/10.1002/pbc.21577>. Published online.
- [54] A.W. van Deutekom, A.J. Lewandowski, Physical activity modification in youth with congenital heart disease: a comprehensive narrative review, *Pediatr. Res.* (2021), <https://doi.org/10.1038/s41390-020-01194-8>. Published online.
- [55] M.C.A. Sprong, W. Broeders, J. van der Net, et al., *Motor Development After Cardiac Surgery in Children With a Congenital Heart Defect: A Systematic Literature Review and Meta-analysis*, Published online, 2020.
- [56] N.H. Amir, D.M. Dorobantu, C.A. Wadey, et al., Exercise training in paediatric congenital heart disease: fit for purpose? *Arch. Dis. Child.* (2022) <https://doi.org/10.1136/archdischild-2020-321390>. Published online.
- [57] M. Gomes-Neto, M.B. Saquetto, C.M. da Silva e Silva, C.S. Conceição, V. O. Carvalho, Impact of exercise training in aerobic capacity and pulmonary function in children and adolescents after congenital heart disease surgery: a systematic review with meta-analysis, *Pediatr. Cardiol.* (2016), <https://doi.org/10.1007/s00246-015-1270-x>. Published online.
- [58] M.L. Morrison, A.J. Sands, C.G. McCusker, et al., Exercise training improves activity in adolescents with congenital heart disease, *Heart* 99 (15) (2013), <https://doi.org/10.1136/heartjnl-2013-303849>.
- [59] P.E. Longmuir, P.N. Tyrrell, M. Corey, G. Faulkner, J.L. Russell, McCrindle BW, Home-based rehabilitation enhances daily physical activity and motor skill in children who have undergone the fontan procedure, *Pediatr. Cardiol.* (2013), <https://doi.org/10.1007/s00246-012-0618-8>. Published online.
- [60] D.M.Y. Brown, D.A. Dudley, J. Cairney, Physical literacy profiles are associated with differences in children's physical activity participation: a latent profile analysis approach, *J. Sci. Med. Sport* (2020), <https://doi.org/10.1016/j.jsams.2020.05.007>. Published online.
- [61] J.J. Noordstar, M.C.A. Sprong, M.G. Sliker, et al., Is measuring physical literacy in school-aged children with cystic fibrosis or congenital heart disease needed? *Pediatr. Phys. Ther.* 35 (1) (2023) <https://doi.org/10.1097/PEP.0000000000000967>.
- [62] B.J. Lentzner, D.M. Connolly, C.K.L. Phoon, Do paediatric cardiologists discuss cardiovascular risk factors with patients and their families? *Cardiol. Young* (2003) <https://doi.org/10.1017/S104795110300115X>. Published online.
- [63] C.A. Williams, L. Gowing, R. Horn, A.G. Stuart, A survey of exercise advice and recommendations in United Kingdom paediatric cardiac clinics, *Cardiol. Young* (2017), <https://doi.org/10.1017/S1047951116002729>. Published online.
- [64] J. Ware, J.L. Butcher, B. Latal, et al., Neurodevelopmental evaluation strategies for children with congenital heart disease aged birth through 5 years: Recommendations from the cardiac neurodevelopmental outcome collaborative, *Cardiol. Young* (2020), <https://doi.org/10.1017/S1047951120003534>. Published online.