10-m Shuttle Ride Test in Youth With Osteogenesis Imperfecta Who Use Wheelchairs: Feasibility, Reproducibility, and Physiological Responses

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Background. Physical fitness levels in youth with osteogenesis imperfecta (OI) who use wheelchairs are unknown. The 10-m Shuttle Ride Test (SRiT) has recently been introduced as a field test to determine cardiorespiratory fitness in children with cerebral palsy who self-propel a wheelchair.

Objective. The purpose of this study was to investigate the feasibility and reproducibility of the SRiT, as well as the physiological responses to the SRiT, in youth with moderate-to-severe OI between 8 and 25 years of age who self-propel a wheelchair at least for long distances.

Design. A test-retest design was used.

Methods. Thirteen patients with OI (8 boys, 5 girls; mean±SD values for age=15.5±6.4 years) using a manual wheelchair performed 2 SRiTs within 2 weeks. Adverse events, reached stage, peak heart rate (HRpeak), peak respiratory exchange ratio (RERpeak), peak oxygen uptake ($\dot{V}O_2$peak), and peak minute ventilation ($\dot{V}E$peak) were the main outcome parameters.

Results and Discussion. All participants performed a maximal effort at both SRiTs (mean±SD values for HRpeak of 195±9 beats per minute [bpm], RERpeak of 1.32±0.16, $\dot{V}O_2$peak of 25.4±5.6 $mL \cdot kg^{-1} \cdot min^{-1}$, and $\dot{V}E$peak of 47.9±18.6 $L \cdot min^{-1}$), without adverse events. The intraclass correlation coefficient of the reached stage showed excellent reliability (.95). Limits of agreement (LoA) analysis revealed acceptable LoA for reached stage (mean bias=−0.58, range=−2.50 to +1.35). There was a low correlation between reached stage and $\dot{V}O_2$peak (r=.61 and r=.45 for the first and second SRiTs, respectively).

Limitations. The influence of wheelchair properties and individually adjusted wheelchair designs was not examined.

Conclusions. The SRiT appears to be a feasible, safe, and reproducible maximal field test in youth with OI using wheelchairs at least for long distances. This field test might be useful to provide an indication of physical fitness and to assess the efficacy of interventions on physical fitness in these patients.

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Osteogenesis imperfecta (OI) is a clinically heterogeneous heritable condition (most often autosomal dominant), characterized by increased bone fragility, predisposing to fractures, low bone mass, bone deformities, and short stature.[1–3] In addition, children with OI frequently exhibit vertebral compressions, blue sclera, premature hearing loss, dentinogenesis imperfecta, easy bruising, hypermobility, low muscle tone, reduced exercise capacity, and reduced muscle force.[1,2,4–6] The exact incidence of OI is unknown; however, it has been estimated at 1 in 10,000 to 20,000 births.[1,7] Patients with OI can be classified according their clinical phenotype, based on clinical and radiographic criteria,[8] as having mild nondeforming (type I), moderate (type IV), severe progressively deforming (type III), or perinatal lethal (type II) OI. Depending on severity, the condition has a great impact on daily life and related functional abilities in patients with OI.[1–3]

Most patients with a severe type of OI, as well as some patients with a moderate type of OI, rely on a manually propelled wheelchair for mobility. Maintaining or even improving autonomous mobility is of great importance for these patients, as they frequently depend on other people in individual and community participation. Wheelchair mobility is influenced not only by mechanical wheelchair properties and individually adjusted wheelchair designs but also by the patient's physical fitness[9] and the environment.[10] When physical fitness is a limiting factor of wheelchair mobility, the assessment of cardiorespiratory fitness is of great clinical relevance, as it is an important marker of overall health in children and adolescents.[11] Moreover, a higher cardiorespiratory fitness in adults is associated with lower mortality.[12,13]

To date, very little work has been undertaken in the field of cardiorespiratory fitness in patients with OI. Our research group was the first to study the physical fitness in ambulatory children with mild-to-moderate OI (type I and type IV).[4,5] We found significantly lower cardiorespiratory fitness levels and muscle force compared with healthy controls. Strikingly, no studies regarding physical fitness are known in patients with OI who self-propel a wheelchair, which might be explained by a lack of suitable tests in these patients for daily clinical practice and a lack of information about the feasibility and safety of performing maximal exercise in these patients. Arm ergometry with respiratory gas analysis is frequently used to assess cardiorespiratory fitness in patients who use wheelchairs; however, this laboratory test requires qualified personnel and sophisticated instrumentation, which often are not available in clinical practice.

Recently, the 10-m Shuttle Ride Test (SRiT) was introduced and investigated as a field test to measure cardiorespiratory fitness in children with cerebral palsy who use wheelchairs.[14] The authors reported the SRiT to be a feasible, reproducible, and valid maximal field test in this pediatric population. In children and adolescents with OI who use wheelchairs, the SRiT also might be useful as a practical field test to assess and evaluate cardiorespiratory fitness over time; however, the SRiT has not yet been examined in this population. Therefore, the aims of this study were: (1) to study the feasibility and reproducibility of the SRiT and (2) to describe the physiological responses to the SRiT in children and adolescents with moderate-to-severe OI who self-propel a wheelchair at least for long distances.

Method

Participants

Children and adolescents diagnosed with OI were eligible to participate when they: (1) were between 8 and 25 years of age, (2) were classified as ≤5 according to the modified Bleck scale (household walker without crutches or cranes),[15] (3) used a manual wheelchair for at least long distances >500 m), and (4) were known at the outpatient clinic of the 2 Dutch OI centers of excellence in the Netherlands (Wilhelmina Children's Hospital of the University Medical Center Utrecht and Isala Clinics, Zwolle). Patients were excluded if their medical status contraindicated exercise or if they were not capable of cooperating with the testing procedures. Informed consent was obtained from the participants (if >12 years of age), as well as from the parents (if participant was <18 years of age). Assent was obtained from each child <12 years of age.

Procedure

Participants attended 2 study visits within a period of 2 weeks (mean±SD values for between-visit time=8.5±4.3 days, range=2–14) at the University Medical Center Utrecht or at Isala Clinics, Zwolle, during which all tests were administered by 2 experienced testers (B.C.B. and E.B.G.R.). Preparation instructions prior to the study visits (eg, restrain from strenuous exercise the day prior to the exercise test)[16] were given to the participants and their parents. Participants used their own manual wheelchair during all tests and were blinded to their performance on each of the tests. Between the study visits, no adjustments were made in wheelchair configuration. At the first study visit, clinical characteristics and muscle force were determined. The SRiT with respiratory gas analysis was performed at both study visits.

Clinical Characteristics

Body mass was determined using an electronic chair scale (Seca 956, Seca GmbH & Co KG, Hamburg, Germany) with a precision of 0.1 kg. Body height and arm span were measured to the nearest centimeter. Body mass index was calculated as body mass divided by the square of the body height. Bone mineral density (with dual-energy x-ray absorptiometry [DEXA] scan findings presented as z scores), fracture history (total number of fractures), presence of intramedullary rodding, presence of scoliosis (Cobb angle >10°), and use of bisphosphonates were extracted from the patient record systems of both hospitals. Clinical characteristics are described in the Results section or presented in Table 1.

Muscle Force

Grip force was measured using a handheld dynamometer (CITEC CT 3001 dynamometer, CIT Technics BV, Groningen, the Netherlands) with the "make" method. The dynamometer was gripped as hard as possible for 3 seconds with the nondominant hand, without pressing the instrument against the body and without touching the elbow to the body. Grip force was measured 3 times. The highest score of the nondominant

Table 1.
Clinical Characteristics of the Participants (n=13)[a]

Participant No.	Sex	Age (y)	Body Mass (kg)	Body Height (cm)	Arm Span (cm)	BMI ($kg \cdot m^{-2}$)	OI Type	Modified Bleck Scale[b]	Wheelchair	BMD L1–L4 (*z* Score)	Fracture History	Sport ($h \cdot wk^{-1}$)	Grip Strength[c] (*z* Score)
1	Female	10.3	35.1	118	125	25.2	III	2	ADL	−1.0	20	1.00	−2.2
2	Female	9.1	18.6	96	104	20.2	III	2	ADL	−0.3	75	1.00	−1.5
3	Male	13.7	46.3	125	156	29.6	III	3	ADL	−2.2	35	1.50	−3.0
4	Female	16.5	89.1	143	171	43.6	IV	1	ADL	−0.6	55	2.50	−0.6
5	Male	11.7	42.2	145	149	20.1	V	4	Sport	−0.9	37	1.00	−3.1
6	Male	10.5	46.2	110	131	38.2	III	2	ADL	NA[d]	100	1.25	−7.7
7	Female	10.0	31.6	120	130	21.9	III	4	ADL	−1.5	10	2.25	−1.0
8	Male	13.2	38.9	134	153	21.7	III	1	ADL	−1.7	250	1.50	−4.1
9	Male	25.6	42.4	110	162	35.0	III	1	ADL	NA[d]	70	1.50	−2.0
10	Female	23.7	36.3	106	151	32.3	III	1	ADL	NA[d]	80	0.00	−1.6
11	Male	9.8	15.0	90	101	18.5	III	2	ADL	−0.6	55	1.00	−8.1
12	Male	24.3	45.4	121	133	31.0	Unknown	2	ADL	NA[d]	7	0.00	−2.5
13	Male	23.2	62.0	153	179	26.5	IV	5	ADL	−2.3	30	1.25	−1.4
$\bar{X}$		15.5	42.2	121	142	28.0				−1.2	63	1.2	−3.0
SD		6.4	18.6	19	24	7.8				0.7	63	0.7	2.4
Range		9.1 to 25.6	15.0 to 89.1	90 to 153	101 to 179	18.5 to 43.6				−0.3 to −2.3	7 to 250	0.0 to 2.5	−0.6 to −8.1

[a] ADL=activities of daily living, BMD=bone mineral density, BMI=body mass index, NA=not applicable, OI=osteogenesis imperfecta.
[b] Modified Bleck scale according to Engelbert et al.[15]
[c] *z* score based on Wind et al.[17]
[d] Undeterminable due to rodding for scoliosis.

hand was compared with normative values for Dutch children and adolescents[17] and expressed as a *z* score (Tab. 1).

Exercise Testing

The SRiT is a field test that might give an indication of cardiorespiratory fitness in children and adolescents who self-propel a manual wheelchair. The test was performed using a detailed version of the protocol developed for children with cerebral palsy who use wheelchairs, as described by Verschuren et al.[14] Methodology of the SRiT in the current study was extensively informed by experiences from their study. As such, a short habituation protocol was completed before starting the SRiT. This habituation protocol consisted of a detailed explanation of the procedure and of the importance of a maximal effort, as well as a short time period to practice the protocol at a low exercise intensity to get used to the procedure and equipment. The habituation protocol was followed by a 5- to 10-minute recovery period before starting the SRiT with a 3-minute rest period to assess baseline cardiopulmonary values.

During the SRiT, participants propelled their wheelchair manually back and forth between 2 lines 10 m apart at a set incremental speed determined by an audio signal, which was played on a standard CD player. Participants started behind one of the lines facing the second line and began riding when instructed by the audio signal. Every completed 10-m track is called a "shuttle." The initial velocity was 2.0 $km \cdot h^{-1}$. Participants continued riding between the 2 lines, turning when signaled by the audio signal. After every minute (stage), the audio signal indicated an increase in speed of 0.25 $km \cdot h^{-1}$. All participants were accompanied and instructed by the same tester during the SRiTs to help them pace themselves with the audio signal. If the 10-m line was reached before the audio signal, the participant had to wait until the audio signal sounded before continuing. The test was finished when the participant reached exhaustion or failed to reach the line (within 2 m) on 2 consecutive audio signals (shuttles), despite strong verbal encouragement. The achieved stage was recorded.

Heart rate (HR) was measured continuously using an HR monitor (Polar T31 transmitter, Polar Electro Oy, Kempele, Finland). Moreover, participants breathed through a firmly fitted face mask, which was connected to a mobile respiratory gas analysis system (Cortex Metamax 3B, Cortex Medical GmbH, Leipzig, Germany). The mobile respiratory gas analysis system was calibrated for respiratory gas analysis measurements (ambient air and a gas mixture of 17% oxygen and 5% carbon dioxide) and volume measurements (3-L syringe) before each test. The metabolic test system consisted of the face mask and a transmitting unit with oxygen and carbon dioxide analyzers carried on the participant's chest (total weight=0.57 kg). The mobile respiratory gas analysis system had a wireless connection with a computer so real-time physical strain of the children during the SRiT could be measured, as indicated by oxygen uptake ($\dot{V}O_2$), carbon dioxide production, respiratory exchange ratio (RER), minute ventilation ($\dot{V}E$), and HR averaged at 10-second intervals (Cortex software version 3.9, Cortex Medical GmbH). The metabolic test system was found to be a reliable and valid system for measuring ventilatory parameters during exercise.[18–20]

Peak $\dot{V}O_2$ ($\dot{V}O_2$peak), peak RER (RERpeak), and peak $\dot{V}E$ ($\dot{V}E$peak), were defined as the average of the last 30 seconds prior to peak exercise (the point at which the SRiT was terminated), whereas peak HR (HRpeak) was defined as the highest value achieved during the last 30 seconds prior to test termination. Subsequently, peak oxygen pulse (O_2 pulse) was calculated by dividing $\dot{V}O_2$peak by HRpeak. The ventilatory threshold was defined as the point at which the ventilatory equivalent for oxygen and the partial end-tidal oxygen tension reached a minimum and thereafter began to rise in a consistent manner, coinciding with an unchanged ventilatory equivalent for carbon dioxide and a peak in the partial end-tidal carbon dioxide tension course.[21,22] When this ventilatory equivalents method appeared to provide uncertain results for a participant's ventilatory threshold, the point at which the linear slope of the relationship between $\dot{V}CO_2$ and $\dot{V}O_2$ changed was taken as the ventilatory threshold, according to the V-slope method.[23] Of the 21 SRiTs with valid respiratory gas analysis data, the ventilatory threshold was determined 8 times (38%) using the ventilatory equivalents method and 12 times (57%) using the V-slope method, whereas the ventilatory threshold was undeterminable once (5%). Furthermore, the $\dot{V}E$-to-$\dot{V}O_2$ relationship ($\dot{V}E/\dot{V}O_2$ slope) up to the ventilatory threshold and the $\dot{V}E$-to-$\dot{V}CO_2$ relationship ($\dot{V}E/\dot{V}CO_2$ slope) up to the respiratory compensation point were calculated. Before and directly after the SRiT, peripheral oxygen saturation (SpO_2) was measured using a pulse oximeter (Nonin Onyx, Nonin Medical Inc, Plymouth, Minnesota) placed on the index finger. Moreover, participants completed a 10-point visual analog scale (VAS) before and directly after the SRiT, indicating their level of overall fatigue.[24] Hereby, the level of perceived exertion of the SRiT (posttest VAS score minus pretest VAS score [ΔVAS]) was assessed.

Data Analysis

Statistical analyses were performed using the Statistical Package for the Social Sciences (IBM SPSS version 22, IBM Corp, Armonk, New York). All data are expressed as mean±SD. Tests for normality were performed on the data using the Shapiro-Wilk test. Except for the ventilatory threshold (%$\dot{V}O_2$peak), O_2 pulse ($mL \cdot beat^{-1}$), and SpO_2 at peak exercise (SpO_2peak), all exercise data were normally distributed.

Feasibility

Descriptive statistics were used to evaluate the feasibility of the SRiT, in which the occurrence of complications and adverse events during or after the SRiT were tracked. Moreover, we evaluated whether participants performed a maximal effort at the SRiT using objective (HRpeak and RERpeak) and subjective (eg, sweating, facial flushing, clear exhaustion) criteria.

Reproducibility

Reproducibility encompasses both reliability and agreement.[25] To assess reliability of the primary outcome measure of the SRiT, the 2-way mixed intraclass correlation coefficient (ICC [2,1]) of the reached stage during the first and second SRiTs was calculated. An ICC >.75 is considered acceptable.[26] For agreement, the standard error of measurement (SEM) was used to determine the precision of the total score of both tests. The SEM assesses response stability, in which it estimates the standard error in a set of repeated scores. The SEM is computed as the standard deviation of the reached stage at the first SRiT multiplied by the square root of 1 minus its reliability coefficient: $SEM = SD \times \sqrt{(1 - ICC)}$. It is important to know, especially in clinical practice, whether the test-retest differences on an individual basis are exceeding the smallest detectable difference (SDD). The SDD of the total score was computed as: $SDD = 1.96 \times \sqrt{(2)} \times SEM$. A Bland-Altman plot[27] was constructed for the reached stage at the first and second SRiTs to check for heteroscedasticity of the test and retest. Furthermore, consistency between the first and second SRiTs was verified graphically. Limits of agreement (LoA) were used to evaluate the level of agreement between the first and second SRiTs. The LoA define the limits within which 95% of the differences are expected to fall (mean±1.96 SD of the differences).

Physiological Responses

To describe the physiological responses to the SRiT (reached stage, HRpeak, RERpeak, $\dot{V}O_2$peak, ventilatory threshold, O_2 pulse, $\dot{V}E$peak, $\dot{V}E/\dot{V}O_2$ slope, $\dot{V}E/\dot{V}CO_2$ slope, SpO_2peak, and ΔVAS), we used descriptive statistics. To test for statistically significant differences between the first and second SRiTs for each physiological variable, a paired-samples *t* test or its nonparametric equivalent, the Wilcoxon signed rank test, was performed, as appropriate. A *P* value <.05 was considered statistically significant.

Role of the Funding Source

This study was supported by an unconditional research grant (R2011061) from the Rehabilitation Fund (Revalidatiefonds, Bunnik, the Netherlands).

Results

In total, 27 children and adolescents between 8 and 25 years of age with OI who use a wheelchair for at least long distances were identified using the electronic patient database of the 2 OI cen-

ters of excellence in our country. Four patients (15%) could not be contacted, and 8 patients (30%) were not willing to participate due to lack of time involvement or safety doubts. Of the 15 patients (56%) who provided informed consent, 13 (48%) were tested. One 15-year-old girl (OI type III) developed severe OI-related back pain between inclusion and the first study visit, and a 13-year-old boy (OI type III) had a hip fracture that did not recover due to low bone mineral density, making it impossible for them to participate. Of the 13 participants who completed the study, 9 (69%) had OI type III, 2 (15%) had OI type IV, 1 (8%) had OI type V, and no definitive (DNA-based) classification was established in 1 participant (8%). All participants had one or more intramedullary rods, all in the lower extremities (tibia or femur, or both), and scoliosis was present in 11 participants (85%). Moreover, all participants had a history of bisphosphonate utilization (pamidronate, zoledronate, or risedronic acid), of whom 10 participants (77%) were using bisphosphonates during the study period. Clinical characteristics of the participants who completed the study are presented in Table 1.

Feasibility

There were no complications or adverse events during or after the SRiTs (up to 2 weeks after the tests). None of the participants complained or showed symptoms of nausea, dizziness, syncope, fainting, or chest pain while performing the SRiT, and they were all capable of performing a maximal effort at both study visits. Mean±SD values for HRpeak and RERpeak attained during the tests were 193±8 bpm (range=180–213) and 1.31±0.19 (range=1.01–1.75), respectively. There were no cases of desaturation (drop of >4% in Spo_2); mean±SD values for Spo_2peak were 98%±1.3% (range=95%–100%). During all SRiTs, participants showed clinical signs of intense effort (eg, sweating, facial flushing, clear exhaustion). The main reasons to terminate the SRiT were arm fatigue (n=25, 96%) and dyspnea on effort (n=1, 4%). The level of perceived exertion during the SRiT, as indicated by the difference between the level of overall fatigue before the test and the level of overall fatigue directly after the test (ΔVAS), was rated as 6.8±1.9 (range=4–10).

Table 2.
Reproducibility Statistics for the 10-Minute Shuttle Ride Test (n=13)[a]

Variable	ICC	95% CI of ICC	SEM	SDD	LoA
Reached stage	.95	.83, .98	0.70	1.9	–2.50 to +1.35

[a] ICC=intraclass correlation coefficient, CI=confidence interval, LoA=limits of agreement, SDD=smallest detectable difference, SEM=standard error of measurement.

Reproducibility

There was no statistically significant difference between the reached stage at the first and second SRiTs. Mean±SD values for the reached stage were 11.9±3.0 (range=6.5–17.0) and 12.5±3.1 (range=8.0–17.5) for the first and second SRiTs, respectively. Table 2 presents the reproducibility statistics of the SRiT. The ICC for the reached stage was .95, in which the 95% confidence interval (CI) ranged from .83 to .98 ($P<.001$), with an SEM of 0.70 and an SDD of 1.9 (16%). As can be appreciated from the Figure, the Bland-Altman plot shows a mean±SD bias between the 2 SRiTs for the reached stage of −0.58±1.00. The LoA values for the reached stage were −2.50 and +1.35, respectively.

Physiological Responses

The physiological responses to the SRiT are represented in Table 3. The SRiT was able to stress all participants maximally (HRpeak >180 bpm and RERpeak >1.0 in all tests). Heart rate increased, on average, about 2 times resting values, from approximately 96 bpm to 193 bpm. The RER values gradually increased, on average, from 0.84 at rest to 1.31 at peak exercise, representing the complete metabolic scope. Oxygen uptake increased, on average, from 271 $mL{\cdot}min^{-1}$ at rest to 1,092 $mL{\cdot}min^{-1}$ at peak exercise, and the ventilatory threshold occurred at approximately 63% of $\dot{V}O_2$peak. The O_2 pulse at peak exercise increased, on average, to 5.8 $mL{\cdot}beat^{-1}$ (12.9 $mL{\cdot}kg^{-1}{\cdot}beat^{-1} \times 100$), and $\dot{V}E$peak was, on average, 47.5 $L{\cdot}min^{-1}$ at peak exercise (1.20 $L{\cdot}kg^{-1}{\cdot}min^{-1}$). Ventilatory efficiency was 21.4 for the $\dot{V}E/\dot{V}O_2$ slope and 25.8 for the $\dot{V}E/\dot{V}CO_2$ slope. There were no statistically significant differences between the first and second SRiTs concerning the reached stage and the physiological responses, except for the attained HRpeak (95% CI=0.13, 8.04; $P=0.04$) and the rating of perceived exertion (95% CI=−2.23, −0.08; $P=.04$). Reached stage correlated poorly with $\dot{V}O_2$peak (first SRiT: $r=.61$, $P=.05$; second SRiT: $r=.45$, $P=.16$).

Sample Size Justification

A justification of the sample size was performed post hoc by using a method to calculate the required number of participants in a reliability study.[28] This approach is based on the number of tests (2 SRiTs), an alpha of .05, a beta of .20, a

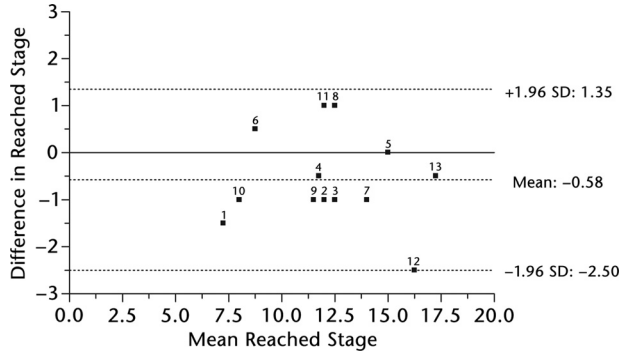

Figure.
Bland-Altman plot of the reached stage during the first and second 10-m Shuttle Ride Tests (SRiTs). A difference in reached stage (y axis) >0 indicates a better test score at the second SRiT, whereas a difference in reached stage <0 represents a better test score at the first SRiT. The numbers above each marker represent participant numbers, which correspond to those in Table 1.

Table 3.
Physiological Responses to the SRiT (n=13)[a]

Variable	First SRiT	Second SRiT	95% CI[b]	*P*	ICC	95% CI of ICC
Reached stage	11.9±3.0	12.5±3.1	−1.18, 0.03	NS	.95	.83, .98
HRpeak (bpm)	196±9	192±9[c]	0.13, 8.04	0.04	.75	.34, .92
RERpeak	1.31±0.15[d]	1.31±0.23[e]	−0.05, 0.08	NS	.89	.63, .97
$\dot{V}O_2$peak (mL·min^{-1})	1,084±387[d]	1,101±408[e]	−75.97, 63.77	NS	.97	.89, .99
$\dot{V}O_2$peak (mL·kg^{-1}·min^{-1})	25.2±5.7[d]	25.2±5.1[e]	−1.56, 1.64	NS	.92	.71, .98
Ventilatory threshold (mL·min^{-1})	711±183[f]	660±189[e]	−18.12, 74.56	NS	.94	.77, .99
Ventilatory threshold (%$\dot{V}O_2$peak)[g]	63±11[f]	62±9[e]	NA[h]	NS	.70	.12, .92
O_2 pulse (mL·beat^{-1})[g]	5.6±2.0[d]	6.2±2.1[i]	NA[h]	NS	.96	.84, .99
$\dot{V}E$peak (L·min^{-1})	47.7±18.6	47.3±18.1	−2.60, 3.53	NS	.96	.88, .99
$\dot{V}E/\dot{V}O_2$ slope	21.0±6.2[d]	21.9±3.5[e]	−6.63, 5.34	NS	−.11	−.69, .56
$\dot{V}E/\dot{V}CO_2$ slope	25.9±4.0[d]	25.7±4.0[e]	−2.94, 3.49	NS	.52	−.17, .87
SpO_2peak (%)[g]	97.8±1.7[j]	98.1±0.9[j]	NA[h]	NS	.41	−.22, .80
ΔVAS	6.2±1.9	7.4±1.7	−2.23, –0.08	0.04	.51	−.03, .82

[a] Values are presented as mean±SD. CI=confidence interval, HRpeak=heart rate at peak exercise, bpm=beats per minute, ICC=intraclass correlation coefficient, NA=not applicable, NS=not statistically significant, O_2 pulse=oxygen pulse, RERpeak=respiratory exchange ratio at peak exercise, SpO_2peak=peripheral measured oxygen saturation at peak exercise, SRiT=10-m Shuttle Ride Test, $\dot{V}E$peak=minute ventilation at peak exercise, $\dot{V}E/\dot{V}CO_2$ slope=minute ventilation-to-carbon dioxide production relationship, $\dot{V}E/\dot{V}O_2$ slope=minute ventilation to oxygen uptake production relationship, $\dot{V}O_2$peak=oxygen uptake at peak exercise, ΔVAS=visual analog scale for the rating of perceived exertion (posttest VAS minus pretest VAS).
[b] 95% confidence interval of the difference between the means.
[c] n=12, heart rate was not determinable in 1 participant.
[d] n=11, invalid respiratory gas analysis measurements in 2 participants.
[e] n=10, invalid respiratory gas analysis measurements in 3 participants.
[f] n=10, invalid respiratory gas analysis measurements in 2 participants, and the ventilatory threshold was not determinable in 1 participant.
[g] Wilcoxon signed rank test.
[h] Not applicable, Wilcoxon signed rank test.
[i] n=9, invalid respiratory gas analysis measurements in 3 participants, and heart rate was not determinable in 1 participant.
[j] n=12, oxygen saturation was not determinable in 1 participant.

minimal ICC of .60, and a maximal ICC of .90. The minimal and maximal ICC values were chosen because an ICC>.75 is generally considered satisfactory.[26] The results indicated that the sample size had to be at least 11.7,[28] or 12 after rounding up.

Discussion

Compared with laboratory tests, the SRiT is a simple and inexpensive field test protocol that provides exercise physiologists or physical therapists working with children and adolescents who manually propel a wheelchair with an indication of the patient's physical fitness. The SRiT requires few resources with respect to specialized equipment (eg, respiratory gas analysis system, arm ergometer) or qualified technicians. Additionally, field tests facilitate a greater involvement and motivation of wheelchair users who are performing in a more familiar environment.[29]

In the current study, 48% of the patients with OI known at the 2 OI centers of excellence in our country who were between 8 and 25 years of age and manually propelled a wheelchair for at least long distances completed all measurements. The results indicate that the SRiT is a feasible, safe, and reproducible maximal field test for children and adolescents with moderate-to-severe OI who self-propel a wheelchair. These findings are in line with the results of a recent study investigating the feasibility, reproducibility, and validity of the SRiT in children and adolescents with spastic cerebral palsy who use wheelchairs.[14] No complications or adverse events occurred during or after the tests in which patients were maximally stressed. In order to evaluate cardiorespiratory fitness, it is essential that the participant performs a maximal effort during an exercise test. For pediatric populations, HR and RER at $\dot{V}O_2$peak are recommended as criteria to assess whether the cardiopulmonary system was stressed maximally.[30] More specifically, an HR at $\dot{V}O_2$peak of at least ≥95% of 195 bpm and an RER at $\dot{V}O_2$peak of at least ≥1.00 are recommended as objective criteria of a maximal effort for cycle ergometry.[30] In the current study, all participants met these criteria and performed a maximal effort on the SRiT. Hence, the SRiT requires a great metabolic demand, during which the cardiorespiratory system is maximally stressed, without any complications or adverse events in these patients.

Furthermore, the results indicate that the SRiT can be performed reproducibly by this population of wheelchair users, as indicated by the high reliability for the reached stage (ICC=.95; 95% CI=.83, .98; $P<.001$), with an SEM of 0.70 and an SDD of 1.9 (16%) (Tab. 2). Agreement

was acceptable (Figure), as reflected by the relatively narrow LoA (−2.50 to +1.35). The average difference between the reached stage attained at the 2 SRiTs was −0.58, indicating that the participants attained, on average, a slightly higher reached stage at the second SRiT (11.9±3.0 and 12.5±3.1 at the first and second SRiTs, respectively; $P>.05$). Next to providing an indication of the child's physical fitness, these results demonstrate that the SRiT also can be used for individual evaluative purposes (eg, within-patient monitoring of physical fitness, evaluating the efficacy of therapy). These findings are in accordance with the study by Verschuren et al,[14] who reported a high ICC for the reached stage (exercise duration) during the SRiT (ICC=.99; 95% CI=.98, 1.00; $P<.001$), with an SEM of 0.50 and an SDD of 1.4, in children and adolescents with cerebral palsy. Their LoA values were −1.45 to +1.45.

Lessons Learned

A low-intensity habituation session prior to the actual SRiT is strongly recommended. By doing this, the patient can get used to the test (eg, the initial speed of the test, the accelerations and decelerations, the turns, the ideal propulsion method). A practice session also accounts for a possible learning effect. In order to ensure a maximal effort of the patient during the SRiT, it is essential to explain the importance of delivering a maximal effort prior to the test, as well as to verbally coach and encourage the patient throughout the test. The monitoring of HR during the SRiT offers the exercise physiologist or physical therapist objective information concerning the delivered effort of the patient and, therefore, is highly recommended.

It is difficult to compare SRiT performance between patients, as wheelchair properties, individually adjusted wheelchair designs, and propulsion skills vary greatly between patients. Vanlandewijck et al[10] reported the impact of different wheelchairs and minimal changes to wheelchair configuration on indoor field test performance. Throughout the SRiT, patients repetitively have to accelerate and decelerate the wheelchair and frequently have to make sharp turns. As a consequence, $\dot{V}O_2$peak prediction from SRiT performance is poor. The reached stage correlated only poorly with the objectively measured $\dot{V}O_2$peak at both SRiTs (first SRiT: $r=.61$, $P=.05$; second SRiT: $r=.45$, $P=.16$). This finding is in agreement with other studies that predicted $\dot{V}O_2$peak from sheer indoor field performance at the 25-m Shuttle Run Test using their manual wheelchair.[31,32]

Hence, strong reservations have to be made concerning the validity of the primary outcome measure of the SRiT (reached stage) as a predictor of $\dot{V}O_2$peak. Instead, the SRiT can be used to quantify within-patient physical fitness longitudinally, provided that the tests are administered in a similar environment and that the patients use their own manual wheelchair without adjustments between the tests. Without respiratory gas analysis, the SRiT provides only a very crude indication of cardiorespiratory fitness, so more sophisticated exercise testing with respiratory gas analysis (eg, arm ergometry, wheelchair-based tests on a treadmill, wheelchair ergometer) may sometimes be required to obtain diagnostic or prognostic information.

Study Limitations

A major limitation of the current study was the fact that the participants were extremely motivated to improve their score achieved at the first study visit when attending the second study visit. Although the participants were blinded to their performance at the first study visit, they were able to recall their reached stage at the first study visit themselves due to the audio signals emitted throughout the SRiT. Furthermore, the influence of the wheelchair properties and individually adjusted wheelchair designs, both important factors in wheelchair mobility,[9,10] was not examined. Together with propulsion skills, these factors might influence SRiT performance. Finally, the study population consisted of a small and relatively heterogeneous group of patients. Osteogenesis imperfecta is a rare disease with several subtypes, and only a small number of patients with OI use a wheelchair. The small sample size in this study precluded further statistical analysis to explore which factors are associated with SRiT performance.

Future Research

Further research is needed to establish the validity and responsiveness of the SRiT as a test to monitor physical fitness in children and adolescents with moderate-to-severe OI who use a manual wheelchair. Important factors such as anthropometry, physiology, environment, propulsion skills, and wheelchair properties should be taken into consideration. Moreover, it would be interesting to evaluate whether these patients benefit from an individual supervised training program to improve physical fitness, as has been shown in children with mild-to-moderate OI.[5] Young adults with more severe types of OI have greater limitations in self-care, mobility, and domestic activities; are less employed; and participate less in sports than young adults with mild involvement.[33] In youth with OI types I and IV, a 12-week individual and supervised physical exercise training program was found to significantly improve aerobic capacity (+18%) and muscle force (+12%) and to reduce levels of subjective fatigue in a safe and effective manner.[5] A higher level of physical fitness in childhood is associated with many health benefits.[11] Moreover, a higher level of physical fitness in patients with moderate-to-severe OI might increase their participation and decrease the restrictions they experience in activities of daily living.

Like the general recommendations for exercise in children and adolescents with mild-to-moderate OI,[34] youth with moderate-to-severe OI should be encouraged to maintain a physically active lifestyle, although contact sports and physical activities with sudden rotation moments of the joints are strongly discouraged. The current study shows that these patients can be maximally stressed in their manual wheelchair without any complications or adverse events. Finally, the SRiT might be useful to evaluate physical fitness at an individual level longitudinally in all childhood diagnoses for children and adolescents who use manual wheelchairs; however, this possibility requires further research. Nonetheless, cognitive and motivational aspects needed to deliver a maximal effort, as well as sufficient skills and motor control required for manually propelling a

wheelchair, are important and might influence SRiT performance.

In conclusion, the SRiT is a feasible and safe maximal field test that can be performed reproducibly in children and adolescents with moderate-to-severe OI who self-propel a wheelchair at least for long distances. This test might be a simple and inexpensive tool to monitor physical fitness and to assess the effect of therapy or physical training on physical fitness in these patients.

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