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Original Article

Activity Intensity During Free-Living Activities in Children and Adolescents With Inherited Arrhythmia Syndromes
Assessment by Combined Accelerometer and Heart Rate Monitor

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Background—International guidelines recommend restriction of activities for many children and adolescents with inherited arrhythmia syndromes to moderate activity (<7 metabolic equivalents [METs]). We hypothesized that moderate levels of intensity would be exceeded during free-living daily activity in these individuals when assessed objectively by combined heart rate and accelerometry monitor (Actiheart).

Methods and Results—Participants wore the Actiheart for ≤7 days on 2 occasions after a maximal exercise test that was used to calibrate the monitor individually against intensity levels. Of 16 participants, 13 (81%) had long QT syndrome, 9 (56%) were female, and median age was 12 years. Monitors were worn for a median (range) of 13 (6–14) days, and a mean (SD) of 11.3 (1.7) hours per day. Vigorous (7 MET) and very vigorous (10 MET) thresholds were exceeded by 15 and 13 participants, respectively. The median (interquartile range), individual, total weekly time spent >7 MET threshold was 113 (65–330) minutes, whereas such time spent >10 MET threshold was 53 (9–115) minutes. Total time >7 MET threshold was 2.3% of monitor wear time. There were no differences in time above threshold between male and female participants (P=0.357) or among those with different levels of activity restriction (P=0.769).

Conclusions—Current recommended activity guidelines are frequently exceeded during routine free-living activities in young participants with inherited arrhythmia syndromes. Whether this indicates increased risk for these individuals or excessively restrictive guidelines remains to be determined. (***Circ Arrhythm Electrophysiol*. 2013;6:939-945.**)

Key Words: arrhythmias, cardiac ■ death, sudden, cardiac ■ exercise ■ pediatrics ■ risk

The inherited arrhythmia syndromes (IAS) are a group of genetically determined conditions responsible for many cases of sudden cardiac death in otherwise healthy young individuals. Exercise and adrenergic stimulation (eg, excitement/anger) may precipitate fainting or sudden cardiac death. The avoidance of intense exercise situations coupled with excitement, which are conditions found during competitive sporting activities and intense play, is recommended.[1,2] International guidelines that outline exercise prohibition for those at risk with IAS lack precision and consistency.[1–4] Leisure time activities with high cardiovascular demand and adrenergic tone may also need to be avoided.[1,2] The overlap between play, daily-living, and sporting activities makes clear distinction of situations that pose risk a challenge in the young.

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Metabolic equivalents (METs) are a standardized method for characterizing intensity of human movement.[5]

Recreational activities judged to be of moderate or low intensity (<7 METs) may be permissible in children and adolescents with IAS.[1] It is not known whether, or how often, these individuals exceed the recommendations during free-living daily activity. Even when competitive sports are avoided, children remain at risk during routine play when intensity can be highly variable. Studies that explore the circumstances of sudden cardiac death in the young show that the minority of events occur during intense competitive activity.[6] Recent investigations have challenged the idea that competitive sports must always be avoided.[7,8]

We hypothesized that objective assessment of activity intensity in children and adolescents with IAS shows instances of activity that exceed the recommended intensity level during free-living daily activity. Heart rates and accelerometer counts calibrated against MET values from an exercise test were the tools we used to determine the intensity and duration of activity during free-living activities.

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Rationale and Methods

Activity Assessment

The accurate measurement of physical activity is complicated in children by the sporadic nature of their activities[9] and is either subjective or objective. Subjective (self-reported) measures are affected by social desirability bias and recall bias.[10,11] Objective measures are either criterion standards (direct observation, indirect calorimetry) or secondary measures (heart rate, accelerometers, pedometers), which must be validated against criterion standards.[12] Accelerometers use piezoelectric transducers to record motion that is converted to count values, which are measured against predetermined cut points.[13] Heart rate increases linearly with exercise intensity and can be distinguished between different levels of activity.[14] For example, moderate-vigorous physical activity in the young is defined as activities producing a heart rate of ≥140 beats/minute; and hard physical activity is correlated with a heart rate of ≥160 beats/minute.[14] Combined accelerometer and heart rate data demonstrate excellent validity when assessed against the criterion standard of calorimetry, and the Actiheart monitor has been validated in children.[15,16]

Participants

Participants were recruited from the Inherited Arrhythmia Clinic at the Children's Hospital of Eastern Ontario. Inclusion criteria were (1) confirmed diagnosis of long QT syndrome (LQTS) or catecholaminergic polymorphic ventricular tachycardia (CPVT), (2) ability to perform a maximal exercise test on the treadmill (7 to 18 years old), and (3) willingness to wear the Actiheart on 2 separate occasions for a period of 7 days each. All participants and parents had undergone routine counseling about competitive sports participation and recreational activity that includes clear identification of the North American (Bethesda Conference #36 [BC#36]) and European recommendations for their specific diagnosis.[3] This counseling explicitly outlines the differences of opinion where they exist, and the rationale for both sets of recommendations. Patients and families are supported to make personal decisions based on their specific circumstances. During follow-up, the level of activity restriction implemented was assessed by parental report. Full restriction was identified as avoidance of competitive and recreational sports activity, and modification/avoidance of full gym class participation at school. Partial restriction was identified as avoidance of competitive sports but involvement in some recreational sports activities and minimal (if any) modification of gym class.

Accelerometer

The Actiheart monitor (CamNTech, United Kingdom) simultaneously records time-stamped accelerometer counts and heart rate via a chest-mounted recorder.[17,18] Output data are counts per epoch and beats per minute for the accelerometer and heart rate data, respectively. Accelerometer counts increase with increasing body movement and intensity. Intensity thresholds for the activity monitors can be determined from values provided from other studies or from individual calibration.[13] Individualized cut points for heart rate are particularly applicable for individuals on β-blocking drugs that alter the usual relationship between heart rate and intensity.

Individual Calibration

We used the Actiheart monitor in a controlled laboratory setting during a formal exercise test to determine individualized cut points for both heart rate and accelerometer count corresponding to moderate (4–7 METs), vigorous (≥7 METs), and very vigorous (≥10 METs) intensities of physical activity.[1,16] On the standard pediatric modified Bruce treadmill protocol, 7 and 10 METs correspond to the end of stage 2 and end of stage 3, respectively.[19]

Study Protocol

Participants were asked to wear the Actiheart for a period of 7 consecutive days on 2 occasions 1 month apart. The Actiheart data are time stamped and permit identification of periods during the day when the heart rate or accelerometer thresholds are exceeded. The participants removed the device during sleep, swimming, and bathing. School days were divided into time at-school and time out-of-school based on parent report.

Participants were asked to complete a physical activity log. Activities were combined into categories such as active transportation (walking, biking), play, organized sports, gym class, school recess, and screen-based activities (eg, video games, television, computer time).

Accelerometer and Heart Rate Event Definitions

Events were defined as (1) heart rate event: the heart rate exceeding the individual threshold, (2) accelerometer event: the accelerometer count exceeding the individual thresholds, and (3) combined event: a contiguous event with onset identified by either the heart rate or accelerometer count exceeding the individual threshold and termination being determined by both heart rate and accelerometer counts falling below the threshold.

Details of data reduction and analysis are given in the Methods section in the online-only Data Supplement.

Statistical Analysis

The outcome variable of interest was duration of events above threshold. Initial data review showed that loss of heart rate data, likely because of skin contact issues, occurred sporadically during many events and, therefore, the analyzed outcome variable for this study was the combined event, as previously defined. Normally distributed continuous variables were described by mean and SD. Nonparametric descriptors (median and range or median and interquartile range [IQR]) were used for non-normally distributed data.

Significance testing used parametric or nonparametric methods as appropriate. Parametric and nonparametric tests for correlated data were used for analysis of event duration data that were correlated because of repeated measurements on individuals.[20] Statistical analysis was performed using Stata 10.1 (College Station, TX). The study was approved by the Research Ethics Board at the Children's Hospital of Eastern Ontario.

Results

Participants, Wear Time, and Threshold Data

Of the 16 individuals who were recruited, 9 (56%) were female, the median age was 12 years, and 81% had a diagnosis of LQTS (Table 1). Family screening identified 6 participants, 5 were asymptomatic probands, and 5 (all 3 of those with CPVT) were symptomatic probands. A personal-use ambulatory external defibrillator was adopted by 8 participants and 2 had implantable defibrillators—1 CPVT patient after cardiac arrest while on β-blockers, and 1 LQTS patient with QTc >500 ms and recurrent syncope while on full β-blockade. Defibrillator lower rates were set at 40 and 80 beats/minute, respectively. BC#36 criteria for restriction were met in 12 of 16 participants. Full restriction of activity was in place in 7 participants, partial restriction in 7, and no restriction in 2 participants. Partial restriction was imposed by the family in 2 individuals who did not meet BC#36 (both with LQTS and QTcs of 460 ms). The mean (SD) QTc in those with LQTS was 476 (31) ms and was longer for those with full restriction compared with partial (505 [33] versus 471 [17] ms; *P*=0.049). All individuals were on β-blockers and 3 with CPVT were also on flecainide. Height, weight, age, and longest QTc did not differ between males and females (Tables 1 and 2).

Table 1.   Descriptive Characteristics of the Study Population

Characteristics	
No. of participants	16
Female, n (%)	9 (56)
Age, median (range)	12 (9–16)
Weight, median (range)	41.1 (23.1–79.9)
Height, median (range)	153.5 (122–182)
LQTS, n (%)	13 (81)
LQTS1	8 (50)
LQTS2	5 (31)
CPVT, n (%)	3 (19)
Activity restriction, n (%)	14 (88)
Full	7 (44)
Partial	7 (44)
None	2 (12)
β-Blockers, n (%)	16 (100)
Nadolol, n (%)	10 (63)
Dose, mg/kg per d, median (range)	1.54 (0.67–2.0)
Atenolol, n (%)	6 (27)
Dose, mg/kg per d, median (range)	1.04 (0.75–1.82)
Flecainide, n (%)	3 (19)
Dose, mg/kg per d, median (range)	2.48 (2.29–4.44)
QTc ms, median (range)	480 (430–550)
Actiheart data	
Days worn, median (range)	13 (6–14)
Hours worn, median (range)	130 (59–182)
Daily school hours, median (range)	12.5 (0.3–15.7)
Daily weekend hours, median (range)	11.6 (1.9–14.9)

CPVT indicates catecholaminergic polymorphic ventricular tachycardia; QTc, Bazett formula–corrected QT interval; and LQTS, long QT syndrome.

All 16 participants wore the device during the first week, and 12 participants provided a separate second week for a total of 184 days of monitoring and a median (range) of 13 (6–14) days. The mean (SD) of total hours of wear time throughout the study per participant was 129.9 (42.2) hours, and the mean (SD) of daily wear time was 11.3 (1.7) hours. There was no difference between daily hours for school days versus weekend days.

The treadmill calibration demonstrated the expected increase in both heart rate and accelerometer counts with increasing workload (Table 2). Compared with normative data, the heart rates were low for the workload level, consistent with

the effects of the β-blockers. There are no sex differences in heart rate or accelerometer counts.

Event Data

7 MET Threshold

In the first week, 15 of 16 participants had events above the threshold, whereas 12 of 12 exceeded this threshold in the second week. The median (IQR) of total time of events above threshold, per participant, was 166 minutes (65–700) during the study period. This equates to a median (IQR) percentage of wear of 2.25% (1.6%–6.9%), which adjusts for different wear times. Median (IQR) daily times above threshold were 21 minutes (7–53), and the median weekly times were 113 minutes (Table 3). There was no statistical difference between weeks 1 and 2 for time spent >7 METs.

10 MET Threshold

This threshold was exceeded by 13 and 10 participants in the first and second weeks, respectively. Daily median (IQR) time >10 METs was 9 minutes (3–29), whereas the median weekly totals was 53 minutes (Table 3). Total time above threshold was higher for week 2 (Skillings–Mack test $P=0.041$).

There were no differences for time spent above thresholds between male and female participants (Wilcoxon rank-sum test $P=0.357$), by level of restriction (Kruskal–Wallis test $P=0.769$), between those with/without an ambulatory external defibrillator at home (Wilcoxon rank-sum test $P=0.753$), those with/without either ambulatory external defibrillator/implantable cardioverter defibrillator ($P=1.0$), those who presented with symptoms ($P=0.126$), or between those who met BC#36 criteria for restriction versus those who did not. Because of the different wear times, these relationships were explored for percentage of wear time above threshold and were not significant for sex (Wilcoxon rank-sum test $P=0.791$) or degree of restriction (Kruskal–Wallis test $P=0.499$).

Activity logs were completed by all participants; however, there was no entry for 35% of all single events. Table 4 shows the amount of time spent >7 MET threshold per week pursuing various activities. There were no differences by sex or the level of restriction. The number of events was insufficient for robust analysis of specific log entries, and descriptive data are provided.

Discussion

This study demonstrates that children and adolescents with IAS frequently exceed the moderate activity levels that are recommended by international guidelines during routine daily

Table 2.   Group Descriptive Statistics of the Threshold Data Obtained During the Individual Calibration Exercise Treadmill Test

	Activity Thresholds (counts/min)			Heart Rate Thresholds (beats/min)			Work
	7 MET (n=16)	10 MET (n=13)	Max MET (n=16)	7 MET (n=16)	10 MET (n=13)	Max HR (n=16)	Max MET (n=16)
25th Percentile	113	189	226	104	123	128	10
Median	141	243	423	112	130	139	11
75th Percentile	175	307	500	119	148	156	12

Data were recorded at the 7 metabolic equivalent (MET) level (all participants), and the 10 MET level if reached (13 participants). The data at the individual maximum level (Max MET) were also recorded (3 of these <10 METs). The data show increasing count levels and heart rates (HRs) as the activity level increases.

Table 3.   Group Descriptive Statistics for Event Duration in Minutes >7 MET and 10 MET Thresholds (by Week and Overall)

	N	25th Percentile	Median	75th Percentile
Minutes >7 METs*				
Week 1	15	66	104	315
Week 2	12	33	205	378
Overall		65	113	330
Minutes >10 METs[†]				
Week 1	13	9	38	76
Week 2	10	12	73	189
Overall		9	53	115

The data show that there are both individual and week-to-week variabilities in the time spent in vigorous or very vigorous activity. METs indicate metabolic equivalents.

*Skillings–Mack test: P=0.366 (week 1 vs week 2).

[†]Skillings–Mack test: P=0.041 (week 1 vs week 2).

and school activities.[1,2] These limits are exceeded across a wide range of daily activities. The cumulative time is substantial and is a call to re-examine our understanding of the guidelines, to reassess beliefs about risk faced by those with IAS during daily activities, and to improve understanding about the manageability of recommendations.

The 3 to 7 MET level that is deemed satisfactory for those with IAS was established without evidence.[1] This level of intensity occurs between 3 and 6 minutes on a modified Bruce treadmill protocol, which is <5th percentile for children >6 years old in recently updated normal values.[21] Recommendations are requiring that healthy children with IAS participate at levels of physical activity of the most unfit of their peers. Our study shows that implementation of instructions limiting activity is difficult during routine family and school life.

All participants met European guidelines for restriction and 12 of 16 participants met BC#36 criteria, but there was variability in how each family described their implementation of these restrictions, from partial to complete restriction. However, there was no difference in the time spent in levels of activity >7 METs between participants whose parents reported full and partial restriction, and is consistent with evidence that self-report is an inaccurate method to assess activity intensity.[10,11] It needs to be emphasized that the majority

Table 4.   Weekly Descriptive Statistics of Event Duration (Minutes) >7 METs for Specific Activities That Were Recorded in the Activity Log Book >10× During the Week

|	Minutes Spent in Each Activity			
	25th Percentile	Median	75th Percentile	Maximum
Sport	11	50	129	294
Transport	13	34	52	579
Play	5	22	35	175
Recess	10	15	58	141
Gym class	9	10	26	108

These data indicate that thresholds are exceeded during a range of activities and not only during sports participation or gym class. METs indicate metabolic equivalents.

of active time was spent <7 METs (the acceptable range), with occasional peaks >7 METs (Figure 1). Many recordings showed mixed patterns of intensity with portions of time spent well above the moderate threshold into the very vigorous range (>10 METs) or above their maximum treadmill level. Our interpretation, which is supported by the activity logs, is that there is substantial variability in how individuals participate in any given activity, and restricting a type of activity, per se, is an inadequate method to restrict intensity of participation in this age group. The self-regulation required for the average child or adolescent to limit the enthusiasm and intensity of their participation in noncompetitive activities to <7 METs may be unrealistic.

Patients with IAS can die during sleep and routine activities.[22] Mitigating the risk to patients during these activities is more challenging than ostensibly decreasing risk by limiting activity intensity. This study raises questions about our understanding of our patients' activities once they leave the clinic. Are the children exceeding the current level of recommended intensity at risk for life-threatening arrhythmias unless we more strictly enforce limitations? Another perspective is to acknowledge our limited understanding of factors that confer risk on a daily basis, and that the level of activity reached in this cohort of patients is not a clear risk factor. Given that the natural history of the IAS is modified by treatment such as β-blockers, the actual safe levels of activity that can be reached is unknown. It is reasonable to conclude that many individuals are regularly performing routine daily activities at vigorous or very vigorous levels without our knowledge. Our assessment of compliance or adherence to our recommendations is flawed as is our understanding of why they have had no events. We may be likely to attribute a good outcome (lack of events) in part to adherence to our advice of limiting activity intensity. If the levels are being exceeded regularly without anyone (physicians, patient, parents) being aware, we are falsely re-enforcing our notion of a successful management strategy.

Alternatively, we could accept the recommendation of activities <7 METs and conclude that our patients engage in high-risk activities without our knowledge. In this case, our response should be to develop better ways to educate ourselves and our patients about self-monitoring and self-regulation of activity intensity. This view would be in concordance with current general practice that is supported by both Bethesda guidelines and the European Society of Cardiology.

Other approaches are not widely implemented at this time. Allowing the option for individuals with LQTS to continue in competitive sporting activities if they choose is an indication of our evolving understanding of the role of activity intensity in risk.[7] There is also substantial variation in application of the guidelines for activity restriction by physicians.[23] At issue is whether or not guidelines should address all diagnoses the same way, or whether there should be diagnosis-specific recommendations. There is some impetus to liberalize the recommendations for LQTS[7]; however, it is still accepted that excessive activity confers high risk for other conditions such as CPVT. Very structured safety plans are initiated for those with LQTS who choose to continue competitive sports, including close supervision with an ambulatory external defibrillator in many cases.[7,8]
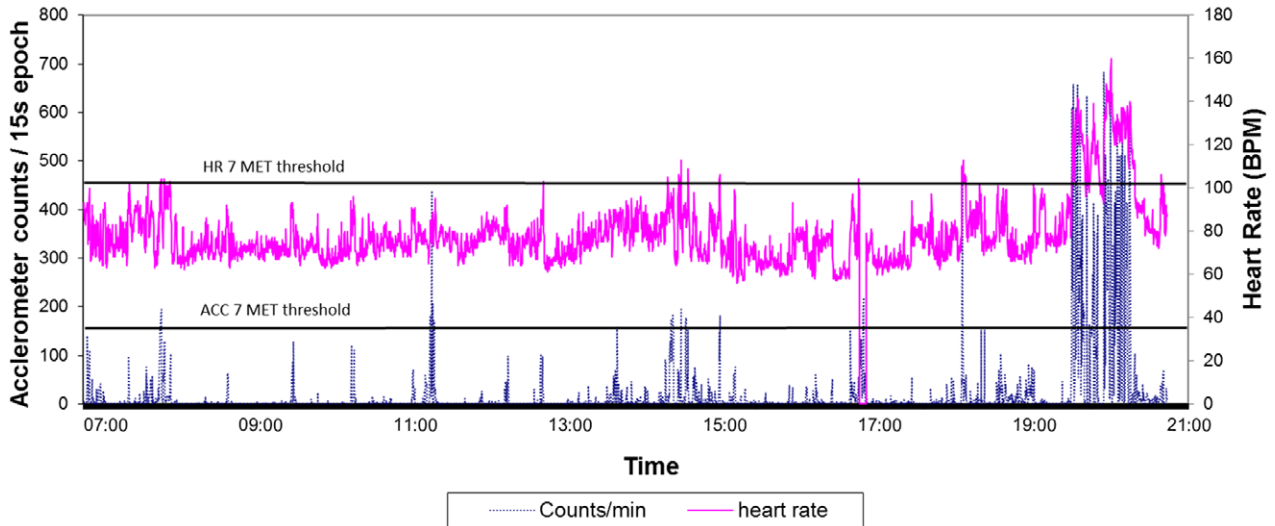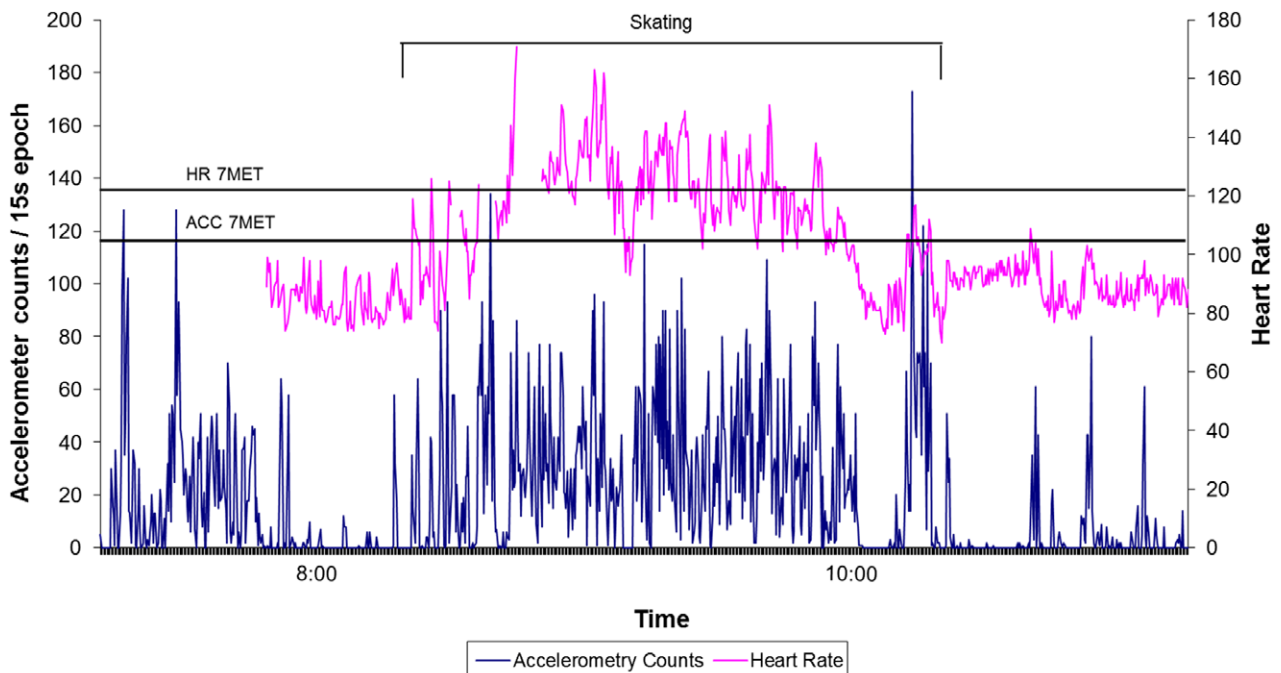
Figure 1. A representative day of a participant wearing the Actiheart device. Accelerometer counts (ACCs) and heart rates (HRs), as well as respective 7 metabolic equivalent (MET) individualized cut points, are provided. The activity shows concordance between the HRs and activity counts.

This is in contrast to the management of the high activity levels during daily activities in this study, many of which are not closely supervised.

Our results highlight the need for a more precise approach to the assessment of activity intensity. There is a rich literature on activity assessment in healthy individuals and tools for objective analysis.[10,12,24] The incorporation of these principles is important to unravel the relationship between activity intensity and outcome in individuals with the IAS, and to inform a reassessment of the limits recommended in the current guidelines.[1] Activity limits that are appropriate to the specific population are important to provide safety

when required, without restricting a healthy, active lifestyle. Objective activity assessment tools will also inform age- and diagnosis-specific recommendations. A compendium of activity intensities relevant to the younger population is available.[25] All physicians who counsel about activity levels and restriction should become familiar with this information, although expansion and further validation of the compendium will be beneficial.

The limitations of this study are important. The participants were selected from a single clinic and did not include inherited cardiomyopathies. The small sample size makes it impossible to draw firm conclusions from detailed analysis

Figure 2. Cross-section of a participant's time spent skating with lower body motion (counts) despite high heart rates (HRs). Accelerometer counts (ACCs) and HRs, as well as respective 7 metabolic equivalent (MET) individualized cut points, are provided.

of associations. Participants were managed by a single physician with a consistent approach to counseling on activity participation. Our cohort of individuals may not be representative of the population of those with IAS in that they may have been much more active or received less adequate counseling about activities to be avoided. The approach to counseling is provided in the online-only Data Supplement. Studies that have broader inclusion criteria and a larger sample size will be required to examine the generalizability of these findings.

The combined heart rate/activity monitor offers advantages, particularly in patients where heart rate control is a therapeutic end point. There was lost heart rate data attributable to skin contact issues during some activity events supporting the selection of a combined event end point. Improving the heart rate data collection would require more permanent and potentially less comfortable skin recording (stickers) rather than the soft strap used. Heart rate and accelerometers capture different types of activities, and those with limited vertical motion (cycling, skating) will show low accelerometer counts with high heart rates (Figure 2). The onset of an event often starts with body motion before the heart rate accelerates and may be better captured by the accelerometer, whereas an isolated increase in heart rate attributable to factors such as emotion will not be accompanied by bodily motion. Given these considerations, the combined monitor offers many benefits for future research.

In conclusion, this study used objective assessment of activity intensity and showed that current recommended activity guidelines are frequently exceeded during routine free-living activities in young participants with IAS. Whether or not this indicates increased risk for these individuals or excessively restrictive guidelines remains to be determined.

Sources of Funding

Disclosures

None.

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CLINICAL PERSPECTIVE

International guidelines recommend restriction of activities for many children and adolescents with inherited arrhythmia syndromes to moderate activity (<7 metabolic equivalents [METs]). These restrictions are frequently applied to competitive sporting activities, and application to free-living activities is problematic. We investigated whether moderate levels of intensity would be exceeded during free-living daily activity in children and adolescents with inherited arrhythmia syndrome when assessed objectively by a combined heart rate and accelerometry monitor (Actiheart). The monitor was worn for ≤7 days on 2 occasions by 16 individuals aged 9 to 16 years, and analysis showed that vigorous (7 MET) and very vigorous (10 MET) thresholds were exceeded by 15 and 13 participants, respectively. The median (interquartile range) individual total weekly time spent above the 7 MET threshold was 113 (65–330) minutes, with 53 (9–115) minutes spent above 10 METS. These levels of intensity were noted during a wide range of activities. This study has shown that current recommended activity guidelines are frequently exceeded during routine free-living activities in young participants with inherited arrhythmia syndrome, and brings to our attention that implementation of instructions limiting activity is difficult during routine family and school life. It remains to be determined whether or not this indicates situations of increased risk for these individuals or excessively restrictive guidelines.

SUPPLEMENTAL MATERIAL

Methods

Accelerometer and Heart Rate Data Reduction

Accelerometer and heart rate data were collected in 15 second epochs and downloaded using the Actiheart software. Auto-cleaning of the data set heart rate points to 0 if they were 1) <30 BPM, or 2) >30 with a rate of change >160 bpm (for 15s epoch). Heart rate gaps of 0 that were less than 5 minutes were interpolated (straight line join), while 0 value gaps greater than 5 minutes were left as 0. Data were then exported to Microsoft Excel (Microsoft, Redmond, Washington, United States), where the number and duration of accelerometer and heart rate events above the individualized threshold values was determined.

The minimum inclusion criterion for each individual's daily recording was at least one hour of continuous data. Events were defined as 1) *Heart Rate Event*: the heart rate exceeding the individual threshold, 2) *Accelerometer Event*: the accelerometer count exceeding the individual thresholds and 3) *Combined Event*: a contiguous event with onset identified by either the heart rate or accelerometer count exceeding the individual threshold and termination being determined by both heart rate and accelerometer counts falling below the threshold. A heart rate event was defined by a rolling average of 15 second epochs totaling 1 minute above the individualized threshold. Below-threshold data points were acceptable within an event if the rolling 1 minute average was above threshold. An accelerometer event was defined by 80% of the epochs being above threshold within a given event. The total time spent above the 7, 10 and maximum-achieved metabolic equivalent thresholds was determined for each participant.

General approach to counselling of patients with diagnosed Inherited Arrhythmia Syndromes

Long QT syndrome (LQTS): The discussions about activity restrictions with children and families of those with LQTS are based on full disclosure of the current recommendations from North America and Europe. Namely, that European Guidelines support restriction from competitive activity for all children with a diagnosis of LQTS, regardless of QTc duration, while North American Guidelines (Bethesda #36) recommend restriction at QTc above 470 msec for males and 480 msec for females[1-3]. The rationale for each approach is discussed, and the fact that these differences in opinion between Europe and North America are an indicator of some uncertainty as to the best approach at this time, and that these will evolve. The risk pyramid is discussed as an introduction to the idea that not all children appear to be at the same risk for events, and activity may be associated with events in some children. Beta blockers are recommended for all children. Children who presented with a symptomatic event or have QTc's greater than gender specific cut-offs are advised strongly to restrict competitive activity and modify participation in gym class; they are encouraged to continue participation in recreational activities that do not involve intense, sustained or burst effort. Asymptomatic children with QTc's below the cut-offs are informed of the North American position that they can participate in competitive activities and the European position of restriction[1-3]. The purpose of the discussions is for patients and their parents to develop an understanding that even children with normal QTc's may be at risk for an event under unpredictable circumstances. It is emphasized that the decision is theirs to make based on our discussions and recommendations. However, even in the "unrestricted" group safety planning is discussed in detail and strongly

recommended. All families are offered a personal use AED; CPR training is recommended for all families, as well as education of school, and assistance developing an individual safety plan at the school if required. Patients are cautioned to avoid dehydration, replenish fluids, make sure they are compliant with their medication and avoid any other medications on the "to avoid list" at www.qtdrugs.org. The activity classification table is discussed as well as its limitations for defining play and recreational activity in children. Parents and children are informed that these are recommendations that are offered in an effort to minimize risk, however, these may change in the future.

Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT): Activity restriction (competitive and intense recreational) is advised for those with CPVT who have ventricular arrhythmias on treadmill testing or Holter. All our CPVT patients are phenotype positive, and receive advice to restrict which is in accord with both European and North American guidelines[1-3].

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