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Participation in thrill-seeking activities by patients with hypertrophic cardiomyopathy: individual preferences, adverse events and physician attitudes.

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Abstract

Background: Thrill-seeking activities are a favorite pastime for people of all ages. Patients with hypertrophic cardiomyopathy (HCM) are often barred from participation on the basis of danger for arrhythmias. Our aim was to collect information regarding the safety of thrill-seeking activities for HCM patients.

Methods: An anonymous online survey invited adult HCM patients to report participation in 11 activities (rollercoaster riding, jet skiing, rafting, bungee jumping, rappelling, paragliding, kayaking/canoeing, motor racing, snowboarding, BASE jumping and skydiving) before and after HCM diagnosis, along with major (ICD shock, syncope) or minor (nausea, dizziness, palpitations, chest pain) adverse events related to participation, and relevant physician advice.

Results: 647 HCM patients completed the survey, with 571 (88.2%) reporting participation in ≥ 1 TSAs (participant age 50.85±14.21, 56.6% female, 8143 post-diagnosis participations). At time of survey, 457 participants (70.6%) were ICD-carriers or had >=1 risk factor for sudden cardiac death. Nine (1.5%) participants reported a major event during or immediately after (60 minutes) of surveyed activity. Minor adverse events were reported by 181 participants (31.6%). In addition, 8 participants reported a major adverse event >60 minutes later but within the same day. Regarding physician advice, of the 213 responders (32.9%) receiving specific advice, 56 (26.2%) were told safety data is absent with no definitive recommendation, while 24 (11.2%) and 93 (43.6%) were told TSAs were respectively safe or dangerous.

Conclusions: In this cohort, participation in thrill-seeking activities rarely caused major adverse events. This information can be used for shared-decision making between providers and patients.

Abstract word count: 249

Keywords: hypertrophic cardiomyopathy, thrill-seeking activities, rollercoaster

Introduction

Hypertrophic Cardiomyopathy (HCM) is the most common inherited cardiomyopathy and an important cause of sudden cardiac death (SCD) in the young[1, 2]. For patients with HCM, intense or burst physical exertion that results in rapid increase in heart rate is considered dangerous and strongly discouraged by society guidelines[3, 4].

Along with athletic activities, thrill-seeking activities (TSAs), especially theme park attractions, are a very popular pastime worldwide, with an estimated 475.8 million theme park visits in 2017 for the top 10 industry groups[5]. These activities also cause rapid and intense heart rate increases through sympathetic activation in the absence of physical exertion[6, 7]. Despite the popularity of TSAs, there is no data on safety for patients with HCM. Participation in TSAs is therefore not discussed in most recent HCM guidelines[3], leaving individual care providers with the burden of determining appropriate guidance.

Addressing the issue of safety for HCM patients wishing to participate in TSAs is challenging. Prospective studies are difficult to design, mainly for ethical reasons, as amusement park rules exclude guests with pre-existing heart conditions from participating. To address these issues, we examined the self-reported participation in TSAs of an international HCM patient cohort, assessing associated adverse events, physician attitudes towards these activities, patient satisfaction and compliance with received advice.

Methods

Adult (>18 years) patients with a diagnosis of HCM were invited to complete an anonymous online survey (https://is.gd/rollercoasterHCM) regarding participation in 11 TSAs: rollercoaster riding, jet skiing, rafting, bungee jumping, rappelling, paragliding, kayaking/canoeing, motor racing, snowboarding, BASE jumping and skydiving. Patients were recruited through patient advocacy organizations, popular HCM social media outlets, email lists and directly from study investigators at the time of clinical visits. Participating HCM expert centers were located in North America, Europe and Australia, with online access free to anyone who wished to be included in the study. Survey questionnaires assessed: 1) patient demographics, 2) HCM-related medical status, including known SCD risk factors and symptoms during everyday activities, 3) participation in TSAs in relation to HCM diagnosis (before and/or after), 4) experienced symptoms and/or adverse events related to participation (nausea, dizziness, palpitations, chest pain, syncope, ICD shock), and 5) physician advice related to participation in TSAs as well as non-TSA athletic activities, along with patient satisfaction with the level of advice on a 5-point Likert scale. The survey remained open between February and July 2018. Study data were collected and managed using REDCap electronic data capture tools hosted at Yale University. The study was approved by Yale's IRB and institutional IRBs at participating centers.

Statistical analyses were performed using IBM SPSS version 24 (Chicago, Illinois). When comparing groups, Pearson's chi squared with continuity correction (for nominal variables) or Mann-Whitney's U test (for ordinal variables) were used. For numerical variables, Student's t-

test (two-tailed) was used. P-values <=0.05 were considered statistically significant.

Results

Demographics of HCM participants

The survey was completed by 647 HCM patients ("responders"), with 571 (88.2%) reporting having participated in ≥ 1 thrill seeking activities ("participants") in their adult life (age at time of survey 50.85±14.21 years). Of those, 229 (40.1%) participated in one or more TSAs before being diagnosed with HCM but not after, 292 (51.1%) participated both before and after diagnosis, and 50 (8.7%) participated only after their HCM diagnosis. The majority of participants were female (326 responders, 56.6%), and residing mostly in North America and Australia [417 (73%) and 99 (17.3%), respectively]. This may be partly attributed to the fact that survey questions and outreach initiatives were in the English language, possibly also reflecting cultural differences and ease of access to TSAs.

TSA participants' demographic data is presented in Table 1.

Table 1. TSA participants' demographics (n=571)

Age		50.85±14.21
Sex	Male	245 (42.9%)
	Female	323 (56.6%)
Race	Asian/Pacific Islander	16 (2.8%)
	Black/African American	7 (1.2%)

	Hispanic/Latino	14 (2.5%)
	White	514 (90%)
	Other	12 (2.1%)
Residence	Africa	1 (0.2%)
	Asia	9 (1.6%)
	Australia	99 (17.3%)
	Europe	24 (4.2%)
	North America	417 (73%)
	South America	2 (0.4%)

Responders were also asked about their medical history and clinical status at the time of survey (not at time of TSA participation). Clinical information on TSA participants is presented in Table 2.

Table 2. TSA participants' clinical information (n=571)*

Age at diagnosis		39.92 ± 15.89
NYHA class†	I	195 (34.1%)
	II	126 (22.1%)
	III	155 (27.1%)
	IV	53 (9.3%)
FHx of SCD		207 (36.3%)
ICD		256 (44.8%)
Past ICD shock		77 (30%)
B-blocker		360 (63%)
Calcium channel blocker		126 (22.1%)
Disopyramide		32 (5.6%)

Anticoagulation		172 (30.1%)
Diuretics		140 (24.5%)
Amiodarone		24 (4.2%)
Genetic testing performed		327 (57.3%)
Pathogenic mutation		176 (30.8%)
Septal reduction therapy	Myectomy	135 (23.6%)
	ASA	36 (6.3%)
LV max thickness		21.05±7.47
Obstructive HCM		201 (35.2%)
LVOT grad		99.92±50.59
Hx of VT		178 (31.2%)
MRI		339 (59.4%)
LGE		31 (9.1%)
Hx of syncope		213 (37.3%)
High risk for SCD [‡]		407 (71.2%)

[*]Data represent patient status at time of survey completion, not time of participation or diagnosis.
[†]Based on symptoms during everyday activities at the time of survey.
[‡]Based on history of ICD, syncope, VT, first degree relative with SCD or LV thickness >=30 mm.

Disease Characteristics

At the time of survey completion, a majority (n=334, 66%) of participants reported being symptomatic (NYHA class 2-4). Approximately one-third of participants (n=201, 35.2%) reported having obstructive HCM based on either general information obtained from their doctor, or a self-reported LVOT gradient > 30 mmHg.

A strikingly high number of responders (n=457, 70.6%) had ICDs or had at least one established risk factor for sudden cardiac death (syncope, ventricular tachycardia, a first-degree relative with SCD or LV thickness > 30 mm)[3]. Nearly half (n=256, 44.8%) of those participating in TSAs reported having an ICD in place.

The most popular TSAs were rollercoaster riding and kayak/canoeing, reported by 500 (87.5%) and 381 (66.7%) patients, respectively. Most patients reported participating in more than one activity, with a total of 8143 post-diagnosis participations overall.

Adverse events associated with thrill-seeking activities participation

Reported adverse events were considered TSA-related if they occurred during or immediately after (within 60 minutes) the activity and potentially related if they occurred on the day of TSA but were temporally unconnected to participation. TSA-related major events (defined as ICD shock or syncope) were rare, with 9 (1.5%) participants reporting syncope and no one reporting ICD shocks during surveyed activity. Mild symptoms associated with TSAs were more common (181 out of 571 patients, 31.6%), with dizziness and palpitations being most often reported (24.9% and 20.7% of participants, respectively).

Regarding potentially related events, 78 participants (13.6%) reported experiencing an adverse symptom or event more than an hour later but within the same day, including 8 major events (2 ICD shocks and 6 syncopal episodes).

Figure 1 provides a breakdown of participant/non-participant numbers as well as major adverse

events related to participation. Table 3 lists symptoms as well as major events reported by participants.

Figure 1. Study responders and major events

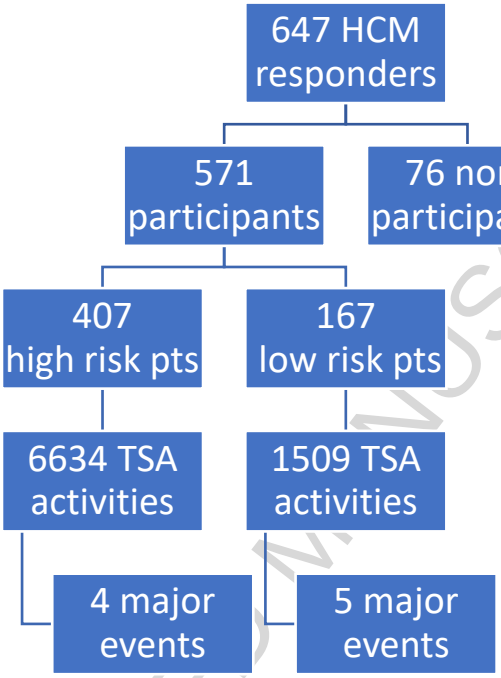


Figure 1 shows the distribution of study responders, high and low risk patients, participations and major events. All major events were syncope.

Table 3. Adverse events and symptoms from participation (n=571)*

	Yes, during or immediately after the activity (within 60 minutes)	Yes, more than an hour later but within the same day
Nausea	64 (11.2%)	12 (2.1%)
Dizziness	142 (24.9%)	31 (5.4%)
Palpitations	118 (20.7%)	39 (6.8%)

Chest pain	59 (10.3%)	33 (5.8%)
Syncope	9 (1.5%)	6 (1.1%)
ICD shock	0 (0%)	2 (0.4%)
Any symptom or event	191 (33.4%)	78 (13.6%)

*Participants could select more than one. Results in table columns are not additive, as participants often reported multiple symptoms.

No discernible clinical pattern was seen for the 9 participants who experienced a major event during a TSA. Table 4 lists their detailed characteristics. They were evenly distributed in terms of gender and presence of high risk features. Three patients did not report heart failure symptoms (NYHA class I), while the remaining 6 were symptomatic at baseline. Age at diagnosis ranged from 14-65 years. The most common trigger activity was rollercoaster riding (4 events), which was also the most popular thrill-seeking activity in the entire participant cohort. Unfortunately, information about the presence of LGE on CMR, a secondary SCD risk factor[8], was not provided by any of the 9 patients. Five out of nine patients reported having participating in TSAs only before being diagnosed with HCM (and not after), while 4 reported participation both before and after their diagnosis. For those 4 patients, the timing of their TSA-related syncopal event in relation to HCM diagnosis (before or after) was not captured.

Table 4. Characteristics of the 9 patients with TSA-related major adverse events*

Patient	A	B	C	D	E	F	G	H	I
Sex	M	F	F	F	F	M	M	M	F
Age	34	57	48	50	47	56	67	66	50
Age at diagnosis	14	39	32	50	31	N/A	65	48	47
Event	Syncope	Syncope	Syncope	Syncope	Syncope	Syncope	Syncope	Syncope	Syncope
Trigger	Racing	N/A	Roller-	Roller-	Roller-	N/A	Roller-	Kayak/-	N/A

activity			coaster	coaster	coaster		coaster	canoe	
NYHA class	III	II	I	IV	I	II	I	II	IV
High risk features †	No	Yes	No	No	No	No	Yes	Yes	Yes
Obstructive HCM	Yes	Yes	No	Yes	No	Yes	N/A	No	Yes
Pathogenic mutation	No test	No test	N/A	Yes	No	No	Yes	No test	No test

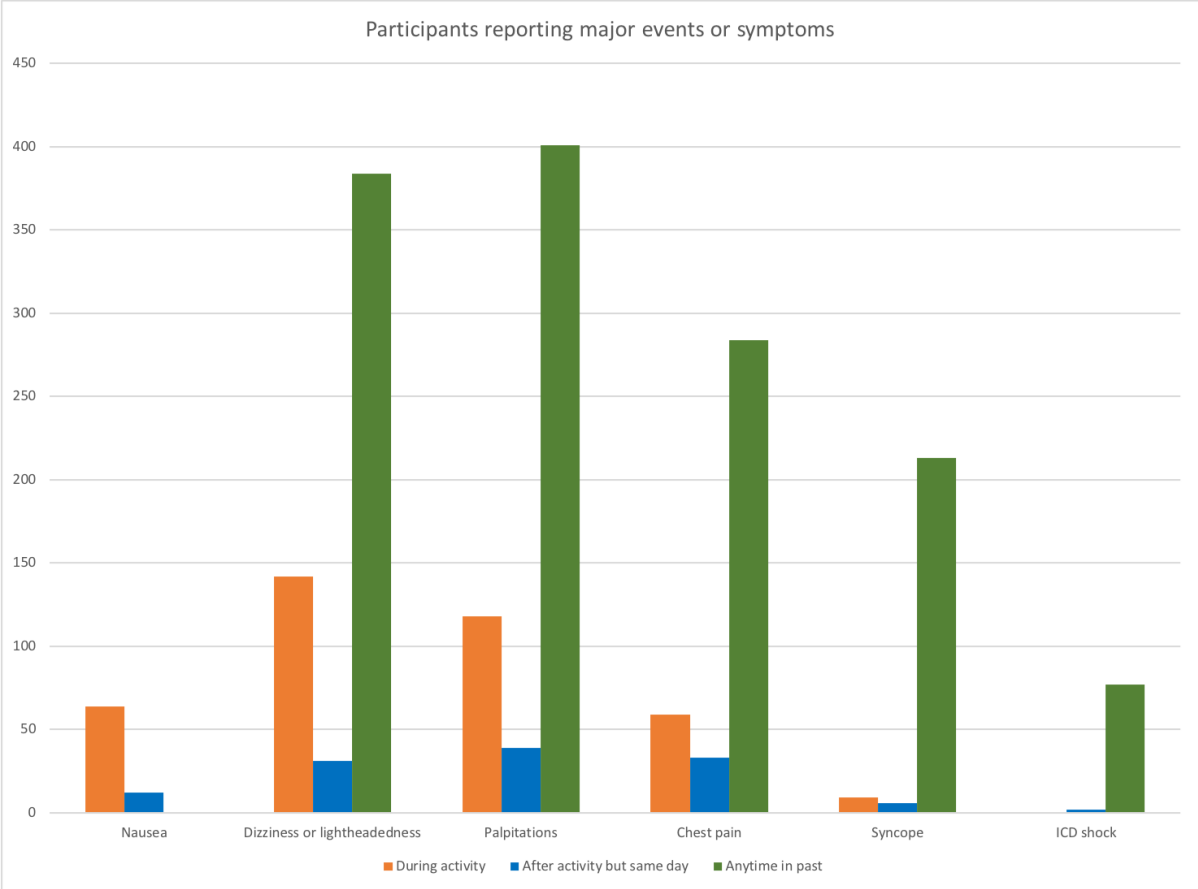
*Three additional participants reported an ICD shock in the survey field reserved for TSA-related events, but when asked to specify the TSA that triggered it they reported non-surveyed activities (hiking, weight lifting, snow skiing). These were added to past ICD shocks in table 2.

†History of ventricular tachycardia (sustained or non-sustained), first degree relative with SCD or LV thickness >=30 mm.

N/A: Not available (patient declined to answer)

Figure 2 provides an overview of the number of participants who reported any adverse symptom or event, either at the time of participation or as part of their medical history.

Figure2.



Number of participants reporting an adverse event, either during TSA participation, on the same day or as part of their medical history.

Physician attitudes and advice provided to patients

Table 5 contains information about physician advice provided to patients regarding participation in TSA or sports activities, as well as patient satisfaction in having their questions answered. All 647 survey responders are included (TSA participants and non-participants). Patients referred to an HCM expert center by their primary care provider/cardiologist were asked to report advice received there, with results stratified according to expert center referral or not.

Table 5. Physician advice and patient satisfaction				
		Referred to expert center (n=429)	**Not referred to expert center* (n=199)**	**p**
Participants		387 (90.2%)	169 (84.9%)	0.152
Non-participants		42 (9.8%)	30 (15.1%)	
Advice provided by physicians on thrill-seeking activity participation	Safe	17 (4%)	7 (3.5%)	0.377
	Dangerous	72 (16.8%)	20 (10.1%)	
	Limited data on safety	40 (9.3%)	16 (8%)	
	No advice	257 (59.9%)	145 (72.9%)	
Advice provided by physicians on sports participation	Avoid all exercise	16 (3.7%)	9 (4.5%)	0.055
	Avoid strenuous exercise but maintain fitness	294 (68.5%)	108 (54.3%)	
	No restrictions on exercise	15 (3.5%)	11 (5.5%)	
	No advice	74 (17.2%)	49 (24.6%)	
Patient satisfaction on received advice (all activities) (5=Very satisfied, 1=Very dissatisfied)		4.1±0.77	3.75±0.66	**<0.001**

*Or did not know/not sure. Responders who did not provide relevant information are not included.

Physician Advice Regarding TSA Participation

In total, 213 responders (32.9%) reported receiving advice specifically on TSAs. Of those, 130 (61%) reported that their doctors spontaneously offered advice on TSAs, while 83 (38.9%) had to specifically ask their doctors about participation. Fifty-six patients (26.2%) were told that safety data is absent for such activities and were therefore given no definitive recommendation,

while 24 (11.2%) and 93 (43.6%) were told that participation was respectively safe or dangerous. The remaining 40 patients replied "Other (I received different advice)" or declined to provide additional information.

We also investigated patient compliance with proffered advice, by examining the subgroup of patients who used to participate in TSAs before their diagnosis and then received specific advice (safe to participate, dangerous or limited safety data) (n=124). Sixteen of the 19 pre-diagnosis participants (84.2%) who were told participation is safe continued participating after their diagnosis, but so did 37 out of the 64 (53.1%) who were told it was dangerous. Of the 41 subjects who were informed that safety data is limited, the majority (35 subjects, 85.3%) decided not to abandon TSAs after their diagnosis.

Physician Advice Regarding non-TSA Athletic Activities

We additionally explored physician advice concerning athletic activities. A majority (n=490, 75.7%) of responders received sports participation advice, whereas 126 (19.4%) received no guidance from their providers regarding non-TSA athletic activities. Fifty-three (10.8%) received extreme guidance by their physicians (complete avoidance of any exertion or complete freedom to participate in any athletics), including 31 patients who reportedly had been referred to an HCM expert center. Having high risk features for SCD did not seem to significantly influence the type of advice provided to participants concerning safety of either thrill-seeking or physical activities.

In general, satisfaction levels with physician advice were high, with 159 responders reporting being very satisfied (score of 5), and only 18 being very dissatisfied (score of 1) with received advice. Patients who had been referred to an HCM expert center were overall more satisfied with their doctor's advice regarding their lifestyle, with an average Likert score of 4.1±0.77 vs. 3.75±0.66 ($p<0.001$).

Discussion

This is the first study to directly investigate safety in thrill seeking activities among HCM patients. The main finding of this study is that in a large cohort of TSA participants, most of whom had at least one high risk feature for SCD, major events (ICD shocks, syncopal episodes) during or immediately after a TSA were rare. Milder symptoms, predominantly dizziness, were commonly reported but did not appear significantly increased compared with the symptoms of daily life. In addition, we found that physicians' advice regarding TSAs was fragmented, even when offered in HCM expert centers, ranging from complete freedom to direct prohibition of participation.

Physiology of TSA Participation

The differential between theoretical concerns based on understanding of physiology and presumed arrhythmic mechanism, and real observed risk is an important region to explore. Based on information available from studies in healthy volunteers, safety concerns for HCM patients who participate in TSAs are not unfounded[6, 7, 9]. Physiological changes from the thrill of participation can be substantial, with burst increases in heart rate despite little or no physical

exertion. Even in TSAs where some physical action is required, such as parachute jumping, heart rate correlates mostly with emotional state rather than exertion, and may reach levels close to or above the age-predicted maximum[10]. Further, though not specifically TSAs, there is an established link between SCD and emotionally intense events such as earthquakes[11] or even while watching sports[12].

This could be concerning for HCM patients, as sympathetic activation, along with tachycardia-induced increased oxygen demands, might be expected to increase atrial or ventricular arrhythmias. Primary physiologic data during TSAs are limited. In three small studies observing a total of 88 healthy volunteers (20 children), continuous ECG monitoring was undertaken during a roller coaster ride[6, 7, 9]. Sinus tachycardia was the predominant rhythm observed, with only one 4-second episode of atrial fibrillation and one 3-beat episode of NSVT recorded.

While our retrospective survey cannot comment directly on arrhythmia prevalence during participation, in a population enriched with ICDs (44%) no shocks were delivered, suggesting a low TSA-induced burden of life-threatening arrhythmias, even in high-risk patients. This is in agreement with a growing body of literature showing overall low arrhythmic propensity in HCM patients during activities that were previously considered stressful[13, 14]. These data suggest that extrapolation of current thinking on physiologic stress response may not accurately predict observed risk.

Mortality

The retrospective nature of this study limits our ability to assess for mortality. Publicly available data for the overall population of theme park visitors suggests that arrhythmias leading to death are uncommon. A study using publicly available information on deaths related to rollercoaster riding reported 7 cases of SCD (including 3 in children) between 1994-2004, with no mention of specific causes[15]. For context, the overall number of rollercoaster deaths during the same decade was 40, and these were primarily caused by traumatic injuries. More recent data from the Saferparks database (www.saferparks.org), covering approximately 8,000 amusement ride accident reports from 28 state agencies and the U.S. Consumer Product Safety Commission (for the years 2011-2016), as well as the National Electronic Injury Surveillance System hospital admissions data (2013-2017) (https://saferparksdata.org/explore-hospital-data), reported a total of 7 cardiac deaths possibly related to amusement rides, with none specifically attributed to pre-existing hypertrophic cardiomyopathy.

The low number of deaths over the span of several years makes significant survival bias unlikely in our study, at least regarding rollercoaster rides. It remains possible that non-reported fatalities from participation in TSAs outside of theme parks exist.

HCM Participants vs General Population

We observed 1.5% of participants with reported major events, all syncopal episodes, during and around the time of TSA. This is similar to the proportion of like events in the general population. The Saferparks database reported 88 loss of consciousness events between 1986-2008 (including seizures) in patrons over 18 years of age visiting U.S amusement parks, accounting for 1.7% of total reported accidents. Interestingly, an additional 8 respondents of our survey reported syncope or ICD shock on the same day as TSA participation, but not during or

within 60 minutes, suggesting that factors other than direct TSA participation may drive symptoms on the day of the event (e.g. hot sun, alcohol intake, fatigue). Other symptoms related to participation, such as dizziness, are also commonly reported by healthy individuals visiting theme park attractions[16].

Physician Attitudes

In addition to identifying adverse events related to TSAs, we also sought to explore physicians' attitudes towards participation, as reflected in advice provided to patients. Impressively, most physicians did not provide any guidance, while a significant portion of those who did were prompted by direct patient queries on the topic. Further, advice specifics were inconsistent, even amongst HCM expert centers. This is not unexpected, as guidance for doctors in published guidelines is lacking, probably due to paucity of data. While current ACC/AHA guidelines grade recreational sports activities according to their acceptability for HCM patients[3], TSAs do not receive direct attention. Older recommendations on inherited cardiomyopathies warned specifically against amusement park rides, but specified that risk was greatest for patients with catecholamine-sensitive or auditory-triggered arrhythmia syndromes such as LQTS and CPVT[17].

Physicians' advice was more uniform concerning participation in sports activities. The majority of doctors advised their HCM patients to avoid strenuous activity as potentially dangerous but encouraged patients to maintain their fitness. This is in accordance with current society guidelines[3]. Importantly, a prospective study is underway to further elucidate the safety profile of physical exertion in HCM patients (LIVE-HCM, ClinicalTrials ID: NCT02549664). A minority of patients reported receiving extreme advice (avoid all physical activity or participate

in sports without restrictions). Extreme advice contradicts available evidence and recommendations, both as regards absolute restriction and permission to participate fully[18].

It is interesting to note that patients referred to expert centers reported being more satisfied, on average, with received advice for participating in TSAs and/or sports. Expert centers did not shy away from giving potentially limiting restrictions (most referred patients were told to avoid high intensity sports and TSAs) so it is clear that satisfaction did not depend on patients getting permissive advice. This, however, contrasts with the low compliance with received advice. Being warned that TSAs were dangerous or that safety data is limited did not deter most patients who enjoyed TSAs before they were diagnosed from continuing to do so. Participating in TSAs is clearly important for a significant number of HCM patients, therefore prospective studies rigorously testing the safety of this behavior may be needed.

Limitations of the study

This study relied on self-reported information and therefore is subject to recall and selection biases. In addition, the possibility of survival bias cannot be excluded; however, we believe a significant effect is unlikely for reasons discussed above. Patient clinical information reflected their status at time of survey completion, not necessarily at time of participation, so any correlations between clinical status and participation in TSAs have to be interpreted cautiously. Some HCM patient races and/or ethnicities were under-represented in the study, as responder population consisted only of English-speaking patients.

The study cohort was highly enriched in patients with risk factors for SCD, as well as patients with advanced heart failure at the time of survey (NYHA class III-IV). This is not a representative sample of the general HCM patient population[5]. We believe this to be partly due to a selection effect, as sicker and/or high-risk HCM patients tend to be more involved with their disease by joining patient advocacy groups, interacting in social media outlets, as well as having more often follow-up clinic visits. This increased the chances of exposure to our survey. An alternative explanation may be that some patients with ICDs felt "protected" from arrhythmias and preferentially participated in TSAs. Regardless of true reason, high-risk patients are expected to be more vulnerable to pro-arrhythmic stimuli and therefore their inclusion in large numbers adds sensitivity to the study.

Conclusions

Thrill-seeking activities are popular amongst HCM patients, despite lack of safety data and often contrary advice from caregivers. We found that participation in thrill-seeking activities was overall safe, with rare major events. Whether these data are sufficiently robust to reassure expert clinician and/or patient concerns based on current thinking about the physiology of stress response in HCM patients remains to be seen. Since this is the first study examining TSAs in the HCM population, it will benefit from prospective validation. In the meantime, providers may consider incorporating this data into discussions with patients at the time of diagnosis and follow-up.

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Conflicts of Interest

None relevant

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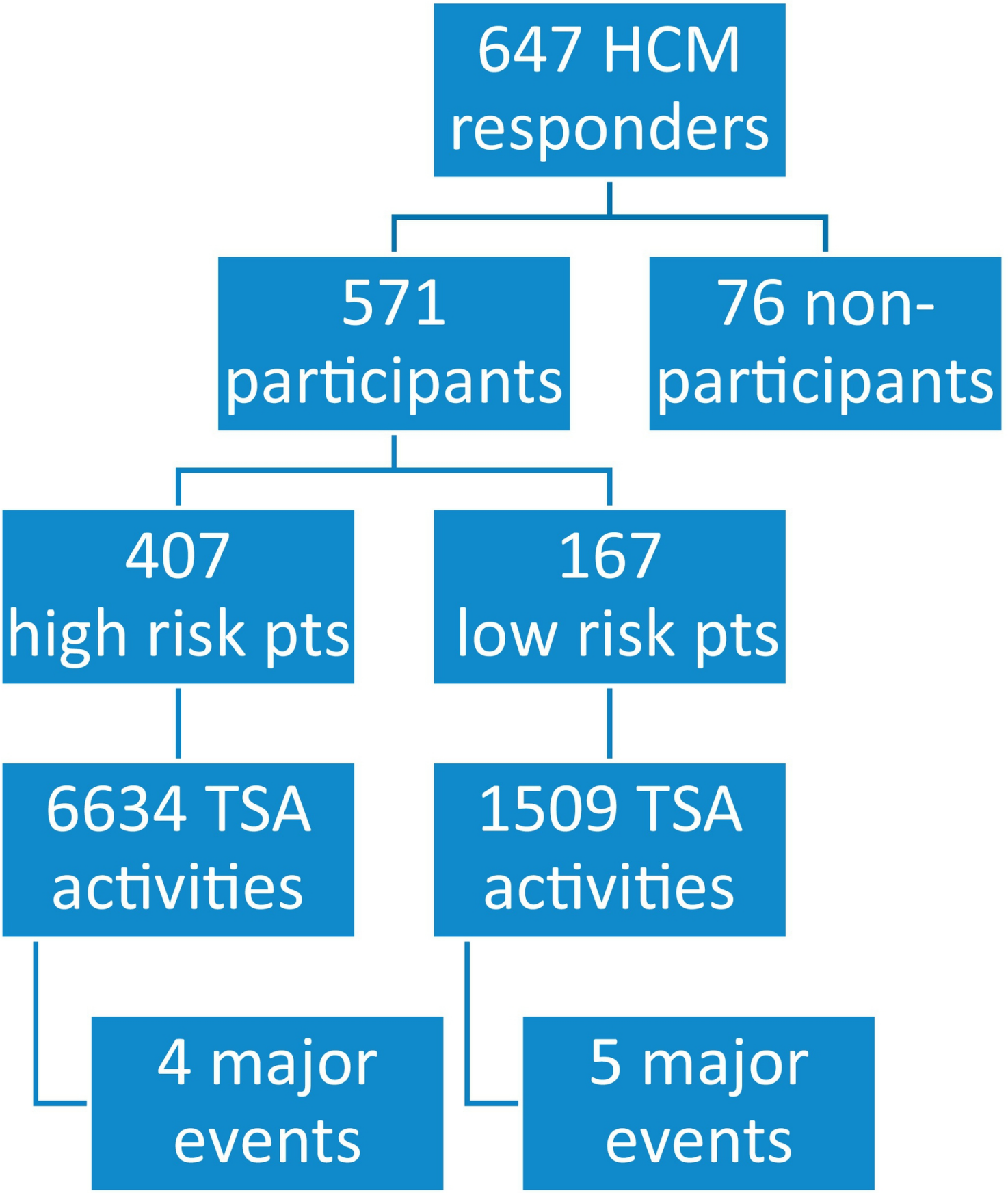
647 HCM responders
571 participants
76 non-participants
407 high risk pts
167 low risk pts
6634 TSA activities
1509 TSA activities
4 major events
5 major events

Figure 1

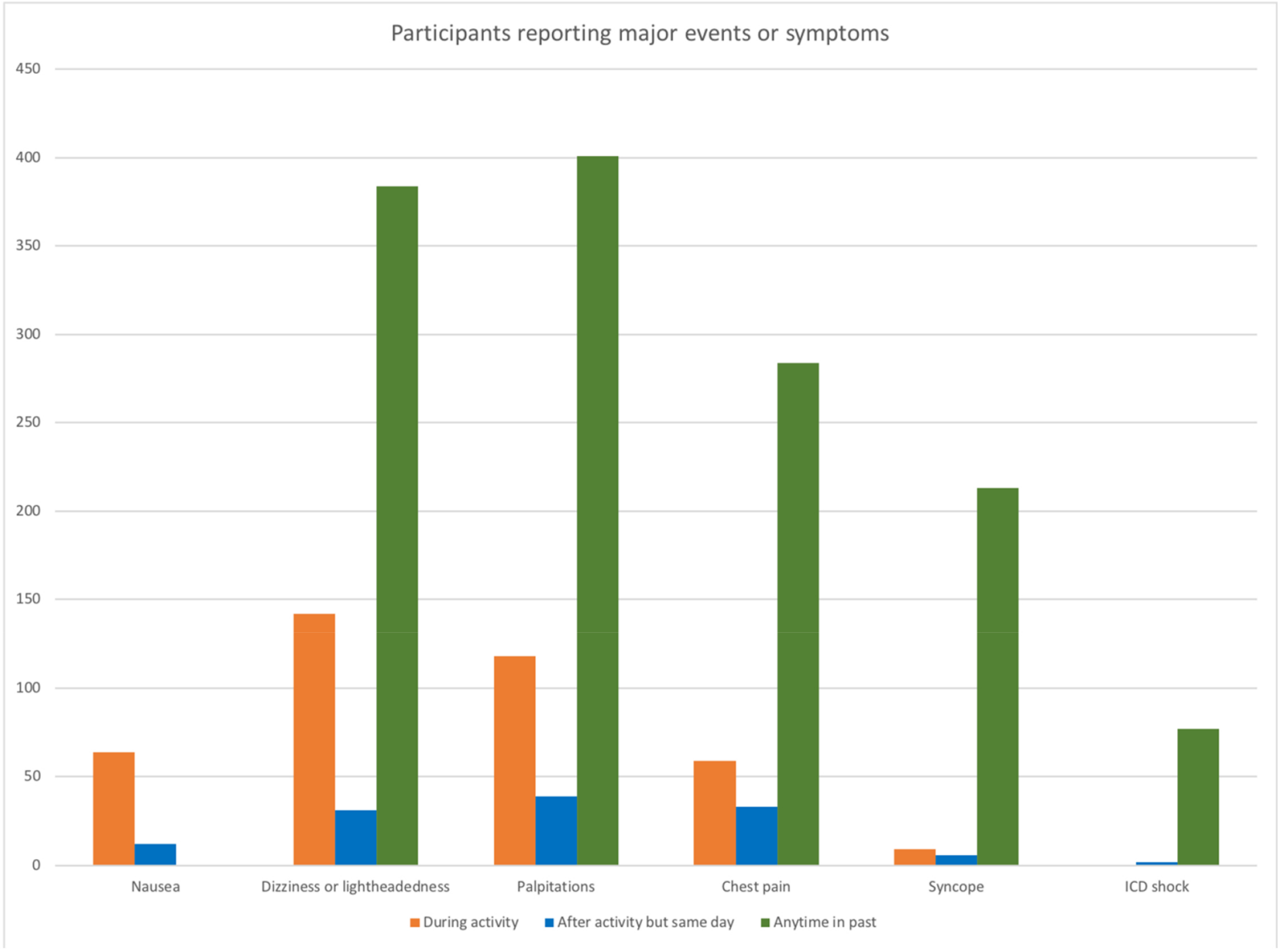
Participants reporting major events or symptoms
450
400
350
300
250
200
150
100
50
0
Nausea
Dizziness or lightheadedness
Palpitations
Chest pain
Syncope
ICD shock
During activity
After activity but same day
Anytime in past

Figure 2