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Regional Implementation of a Pediatric Cardiology Syncope Algorithm Using Standardized Clinical Assessment and Management Plans (SCAMPS) Methodology

Yvonne Paris, MD; Olga H. Toro-Salazar, MD; Naomi S. Gauthier, MD; Kathleen M. Rotondo, MD; Lucy Arnold, MD; Rose Hamershock, MA; David E. Saudek, MD; David R. Fulton, MD; Ashley Renaud, RN; Mark E. Alexander, MD; on behalf of the New England Congenital Cardiology Association (NECCA)*

Background—Pediatric syncope is common. Cardiac causes are rarely found. We describe and assess a pragmatic approach to these patients first seen by a pediatric cardiologist in the New England region, using Standardized Clinical Assessment and Management Plans (SCAMPS).

Methods and Results—Ambulatory patients aged 7 to 21 years initially seen for syncope at participating New England Congenital Cardiology Association practices over a 2.5-year period were evaluated using a SCAMP. Findings were iteratively analyzed and the care pathway was revised. The vast majority (85%) of the 1254 patients had typical syncope. A minority had exercise-related or more problematic symptoms. Guideline-defined testing identified one patient with cardiac syncope. Syncope Severity Scores correlated well between physician and patient perceived symptoms. Orthostatic vital signs were of limited use. Largely incidental findings were seen in 10% of ECGs and 11% of echocardiograms. The 10% returning for follow-up, by design, reported more significant symptoms, but did not have newly recognized cardiac disease. Iterative analysis helped refine the approach.

Conclusions—SCAMP methodology confirmed that the vast majority of children referred to the outpatient pediatric cardiology setting had typical low-severity neurally mediated syncope that could be effectively evaluated in a single visit using minimal resources. A simple scoring system can help triage patients into treatment categories. Prespecified criteria permitted the effective diagnosis of the single patient with a clear cardiac etiology. Patients with higher syncope scores still have a very low risk of cardiac disease, but may warrant attention. (*J Am Heart Assoc.* 2016;5:e002931 doi: 10.1161/JAHA.115.002931)

Key Words: adolescence • ambulatory care • pediatrics • syncope (fainting)

Neurally mediated syncope—an abrupt, reflex-mediated transient loss of consciousness—is very common with a cumulative incidence of 35% by age 18 and nearly 50% by age 21.¹ Newly recognized cardiac disease is rare. While not all pediatric patients with syncope come to medical attention, the mismatch of an exceptionally common cardinal symptom

and a rare but potentially life-threatening cause motivates frequent referral and cardiac evaluation.

The evaluation of these patients can be extensive, expending considerable time and resources.^{2–4} Previous reports on syncope have been mostly observational.^{4–6} To address this, a group of pediatric cardiologists from around

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*Accompanying Appendices S1, S2 and list of additional participants in the New England Congenital Cardiology Association (NECCA) are available at <http://jaha.ahajournals.org/content/5/2/e002931/suppl/DC1>

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New England collaborated to design and implement a quality improvement tool utilizing Standardized Clinical Assessment and Management Plans (SCAMPs) to guide the care of patients presenting to a pediatric cardiology outpatient clinic with a complaint of syncope. SCAMPs methodology uses prospective acquisition of defined data that are analyzed on a recurring basis. A sound initial approach is combined with specific, testable, targeted data statements about either diagnostic or therapeutic approaches. Changes to the care pathways are based on this analysis.^{7,8}

New England Congenital Cardiology Association (NECCA), an organization of ≈ 16 academic and community-based practices representing all 6 New England states, previously successfully implemented a SCAMP on chest pain resulting in a decrease in practice variation and resource utilization.^{7,9,10}

The syncope SCAMP objective was to develop a standardized approach to evaluation of syncope in the outpatient setting. The pathway which emphasized the critical aspects of history, focused family history, physical exam, orthostatic vital signs, and an ECG was asserted to have a high negative predictive value. This would allow identification of the smaller number of patients with higher probability of significant cardiac disease or inherited arrhythmia syndromes and limit the more complex testing. Once the cause of syncope was identified, specific approaches to management were outlined with an emphasis on education and nonpharmacologic management for patients with typical syncope.

Methods

We developed a SCAMP clinical care pathway for syncope using history, physical examination, and ECG.^{11,12} This evaluation targeted pediatric, adolescent, and young adult patients 7 to 21 years of age who present to pediatric cardiology outpatient practices for a chief complaint of syncope. The SCAMP was designed to explore multiple questions simultaneously by targeted data statements. These formed the basis for the clinical care pathway and SCAMP Data Form (SDF) used to track the SCAMP. The strategies for implementation were shared and the guideline was revised based on feedback. Flexibility in implementation of the SCAMP was necessary due to the variation of resources available at different sites. Specific pathways were outlined for typical, exertional, convulsive, atypical/refractory syncope, and postural tachycardia syndrome (POTS) (Appendix S1).

This care pathway was specifically designed to classify the type of syncope and to identify cardiac causes of syncope using the resources in the outpatient cardiology clinical setting. The intent was to complete the evaluation in a single visit for patients with typical, low-severity syncope.

The institutional review boards at Boston Children's Hospital and the participating NECCA sites evaluated the project and waived review designating the SCAMP methodology as a quality improvement initiative. Data use agreements limited reporting age/sex from NECCA sites.

All patient data were de-identified. Activity at the NECCA sites was facilitated, tracked, and coordinated through monthly conference calls that ensured uniformity in use of the SCAMP. Following a pilot period, minor revisions were made and the SCAMP was deployed throughout NECCA for 15 months followed by data analysis and revision of the process with continued tracking for 18 months. Practices entered the SCAMP at different times, so that not all practices were involved the entire 33 months.

There were 2 phases of analysis. Phase 1 used the initial clinical pathways with interim analyses that motivated modification to the revised Phase 2 pathway. Phase 1 analysis included patients enrolled between March 2012 and July 2013, with Phase 2 analysis representing patients entered between August 2013 and December 2014. Interim analyses were also performed. Each analysis was presented broadly within NECCA for feedback followed by modification of the algorithm by consensus.

Patient Selection

Ambulatory patients between 7 and 21 years of age presenting to an outpatient pediatric cardiology practice for a first-time evaluation of the principal complaint of syncope were enrolled. A complete history and physical exam were performed and an ECG was reviewed.

Patients were excluded for cardiac arrest needing sustained cardiopulmonary resuscitation, previously diagnosed seizure, or known cardiac disease. Those with WPW (Wolff Parkinson White) or QT interval >480 ms (Long QT Syndrome) on initial ECG were also excluded. The SDF did not track exclusions.

Data Collection

Clinical evaluation and classifications

Based on initial history and exam, each patient was classified as having typical, exertional, convulsive, atypical/refractory syncope, or POTS. Clinical workflow was completely at the clinician's discretion, such that echocardiogram or other testing could be scheduled prior to the history and physical.

Demographics and clinical characteristics were collected including signs, symptoms and consequences of the syncope event as well as pertinent past medical and family history. Historical data were also noted with regard to past evaluations for syncope and previous specialist or emergency room evaluation. Pertinent physical exam findings,

ECG interpretation, and orthostatic vital signs were recorded either manually or using automated equipment. Orthostatic blood pressure measurements were discontinued during Phase 2 based on analysis of Phase 1 data. The patient judged their symptom severity using 2 subjective, 10-point syncope scales for daily dizziness and overall well-being/wellness. The provider assessed a 0 to 12 point Syncope Severity Score (SSS) based on frequency, severity, and disability associated with the symptoms (Appendix S2).

The initial ECG was evaluated and any abnormalities were noted. Pertinent abnormalities were tabulated, but specific criteria for each ECG diagnosis were neither mandated nor independently reviewed. Echocardiograms were recommended for predefined, generally accepted consensus-based criteria focusing on exertional symptoms or concerning features of the presentation (abrupt onset, injury), abnormal ECG or examination, and potentially concerning family history. Echocardiograms done for family/clinician anxiety were tracked. Patients defined as having typical low-severity syncope were provided with teaching materials and reassurance and not scheduled for follow-up. A complete blood count and ferritin level were recommended for those defined with moderate to severe typical syncope.^{13,14} Provider preference regarding medical treatment or conservative management was recorded, and patient follow-up in 2 months to review response to treatment was recommended.

If patients reported a syncopal event with or immediately following exercise, they were classified as having exertional syncope. Echocardiograms were recommended for those patients who were noted to have syncope during exercise. If no abnormalities were noted on the echocardiogram, then treadmill exercise testing was recommended. Patients exited the SCAMP care pathway if either a structural abnormality or a clinically important arrhythmia were identified. Otherwise, management mirrored typical syncope.

Syncope was classified as convulsive if the patient was noted to stiffen or have brief tonic-clonic movements during the event. Repeated episodes, sustained tonic-clonic events, or neurological findings dictated neurology referral. Repeated episodes directed an exercise test and an echocardiogram as well as ambulatory arrhythmia testing with a loop recorder if the frequency of convulsive syncope was greater than twice a month.

POTS was initially defined as a change in heart rate (HR) of >30 beats per minute during the orthostatic testing process delineated in the SCAMP care pathway with plausible symptoms.¹⁵ Atypical syncope was defined as SSS >8 with combinations of disabling symptoms, syncope while supine, delayed recovery, or sustained “jerking/stiffening” combined with frequent events. More expansive laboratory screening and monitoring was recommended with SSS >4 with particular focus on those with SSS \geq 8.

Treatment with iron was suggested if ferritin levels were <50 ng/mL. Patients with endocrinopathies were referred and exited the SCAMP. Provider preference regarding medical treatment or conservative management was recorded. Patients were recommended for follow-up in 2 months to review response to treatment if the patient had moderate or more severe syncope.

Test interpretation

ECG and ancillary testing interpretations were tabulated based on results recorded on the SDFs. Testing was generally obtained at the time of the initial visit or soon after. Test results performed prior to the pediatric cardiology visit were also noted. There was no attempt to validate results. Ambiguity on critical data was resolved by querying the submitting clinical site.

SCAMP data collection and analysis

Data were collected on an SDF completed by the provider at the initial visit (Appendix S2). Patients who had scheduled follow-up or who returned after discharge from the clinic were evaluated using a second SDF documenting interval history self-reports/assigned SSS. Data were collated by the Institute for Relevant Clinical Data Analytics for NECCA and operations staff at Boston Children’s Hospital.

Results of testing were tracked to assess adherence to the recommendations of the algorithm and to analyze outcomes.

Statistical Analysis

Summary statistics were tabulated. Differences in patient and family history by site and syncope type were examined with χ^2 and Fisher exact tests. Chi-square and Fisher exact tests were also used to analyze the difference in ECG results over syncope types. Bubble plots and correlation analysis were performed on the relationship between physician-assigned and patient-assessment scores. Logistic regression analyzed differential medication use. Lastly, the receiver operating characteristic c-statistic was calculated to determine whether the differences in orthostatic supine values and 3-minute standing orthostatic values were useful in determining syncope classifications. With the exception of specific analyses of follow-up, all analyses were performed on the initial episode of care to maintain independence.

Results

Patient Population

Between March of 2012 and December of 2014 a total of 1292 patients (1437 total visits) were enrolled with 91%

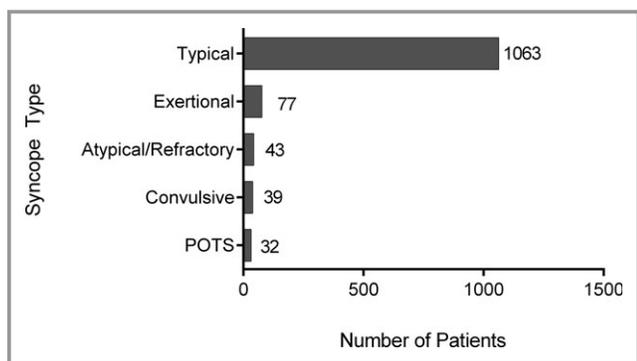


Figure 1. Patients enrolled by syncope type. Distribution of patients enrolled throughout the SCAMP by clinician-assigned syncope type. POTS indicates postural tachycardia syndrome; SCAMP, Standardized Clinical Assessment and Management Plan.

having a single visit. Of those initial visits, 38 were poorly characterized and excluded. Of the remaining 1254 patients, 1063 (85%) were classified as having typical syncope with the remaining 191 (15%) being classified as atypical/refractory 43 (3%), exertional 77 (6%), convulsive 39 (3%), or POTS 32 (3%) (Figure 1). Of the 1254 initial visits, 836 or (67%) were evaluated at Boston Children's Hospital and the remaining 418 were seen at NECCA sites. The distribution between sites was comparable ($\chi^2 P=0.08$).

For the Boston Children's Hospital cohort, 63% were female with 462 (88%) of the females classified as having typical syncope compared to 83% of the males (Table 1). The average age was 14.5 ± 3 years with age-expected heights and weights associated with a mean body mass index of

21 ± 4 . There was a trend toward proportionally more males with syncope not classified as typical, though this did not reach significance. Because typical syncope was diagnosed in the vast majority of patients, this category became the primary focus of subsequent analysis.

Initial Evaluation

History and physical exam

Historical details are outlined in Table 2, which represents positive responses to the questions. Over 80% had dizziness or lightheadedness and experienced a prodrome. Typically 2% to 5% of all questions had missing data.

By design, the exercise questions selected patients with exertional syncope, but otherwise responses did not vary with syncope type. The questions did not effectively capture triggering events other than exercise. Notable trends in the history were relatively frequent palpitations (6–28% with increased frequency in atypical/refractory and POTS patients). By definition, there were fewer school absences in those with typical low-severity or exertional syncope.

The family history was negative for cardiac disease in 1039 or 83% of all patients. Pertinent positives were noted in a significant minority (Table 2). There were significantly lower rates of a family history of sudden deaths for patients with typical syncope compared to all other patients (Fisher's $P=0.02$) Otherwise, there were no significant differences between those with typical and other than typical syncope.

At the time of the initial cardiology evaluation, 284 (23%) had been seen by another specialist. Seventy-seven percent

Table 1. Demographics by Syncope Type

	Total	Mild Typical	>Mild Typical	Atypical/Refractory	Exertional	Convulsive	POTS
Age, y*							
N	836	600	123	23	48	20	22
Mean (SD)	14.5 (3.0)	14.1 (3.1)	15.7 (2.2)	15.6 (2.4)	15.2 (1.9)	14.5 (2.8)	14.5 (2.4)
Sex*							
N	836	600	123	23	48	20	22
Male, n (%)	313	237 (76)	24 (8)	13 (4)	16 (5)	11 (4)	12 (4)
Female, n (%)	523	363 (69)	99 (19)	10 (2)	32 (6)	9 (2)	10 (2)
BMI, kg/m ²							
N	1122	782	165	40	71	35	29
Mean (SD)	21.4 (4.4)	21.2 (4.4)	21.6 (4.2)	23.3 (4.0)	21.6 (4.0)	21.7 (5.5)	21.3 (4.0)
SSS							
N	1249	870	188	43	77	39	32
Mean (SD)	2.5 (1.9)	1.8 (1.1)	5.3 (1.4)	4.3 (2.9)	2.1 (1.6)	2.9 (2.3)	5.1 (2.9)

BMI indicates body mass index; NECCA, New England Congenital Cardiology Association; POTS, postural tachycardia syndrome; SSS, Syncope Severity Score.

*Data-use agreements limited age and sex data from NECCA sites.

Table 2. Patient and Family History Characteristics

Characteristic	n (%)
Patient history	
Patient experienced prodrome?	1080 (89)
Dizziness or light headedness?	1060 (86)
Syncope with rest?	944 (76)
Symptoms resulted in a syncopal event?*	521 (76)
Visual change?	853 (70)
Patient experienced color change?*	604 (60)
Patient experienced loss of tone?*	661 (59)
Anxiety?	276 (23)
Patient incurred injury during the event?	215 (17)
Syncope postexercise?	179 (14)
Palpitations?	129 (11)
Absences from school?	148 (12)
Patient had convulsive-like activity?	107 (9)
Syncope with exercise?	106 (9)
Palpitations unrelated to prodrome?	98 (8)
Patient experienced incontinence?	35 (3)
Was there any level of disability with the event?	78 (7)
History of recurrent joint dislocation?	22 (2)
Family history	
Frequent fainting (first-degree only)?	199 (17)
Arrhythmia/pacemaker/ICD?	116 (10)
Congenital heart disease?	68 (6)
Cardiomyopathy?	30 (3)
Sudden unexplained death (<50 years)?	30 (2)
Heart failure (<50 years)?	17 (1)

Individual patients could report multiple history findings. The presence of color change and loss of tone resulted in unknown or missing responses in >10% of SDFs. One question “Did the patient experience a syncopal event?” was not answered by 46% of respondents and was judged unreliable. ICD, implantable cardioverter defibrillator; SDFs, SCAMP Data Forms.

*≥10% missing.

of these patients were classified as having typical syncope. Patients with atypical or convulsive syncope and POTS were significantly more likely to have seen other specialists (40% to 60%). Exertional syncope patients were least likely to have been referred to another specialist at 12% (overall $P<0.01$). Overall, prior emergency room visits had occurred in 40% of patients. Those with typical syncope were less likely to have had prior emergency care ($P<0.01$).

A significant minority of patients (39%) reported medication use. Those with a SSS ≤ 4 had a lower proportion of medication use than those with a higher score (35% versus 67%) with higher SSS strongly associated with increased medication use (Odds Ratio: 1.395%, CI: 1.2–1.4 for each

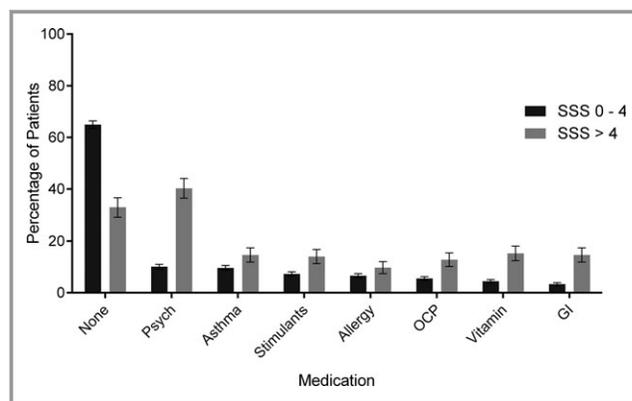


Figure 2. Medication use by syncope severity. Bar graph of medication use is stratified by syncope severity. The majority of patients with typical low-severity syncope and exertional syncope were not on any medications, with 33% of typical low-severity patients on a wide range of medications, none with a frequency >10%. More symptomatic patients were frequently on medications, with that finding most apparent for psychiatric medications, and statistically significant with several other medication classes. Increased Syncope Severity Score (SSS) was strongly associated with increased medication use (odds ratio: 1.3, 95% CI 1.223–1.4, per 1-point increase in syncope score, $P<0.0001$). GI indicates gastrointestinal; OCP, oral contraceptive.

1-point increase in syncope score $P<0.0001$) (Figure 2). When stratifying by syncope type, patients with typical low-severity syncope and exertional syncope had the lowest reported medication use. This finding was most prominent with psychiatric medications, though true for several other classes of medication.

Syncope scores

There was the expected asymmetric distribution of syncope scores, with 79% of patients reporting overall wellness scores >7 (10 “perfect”), and 63% dizziness scores <3 (10 “falling down/dizzy every time I stand”). Physician-assigned score was similarly <3 in 58% indicating minimal severity. Patients with atypical syncope and POTS had higher dizziness scores, lower overall wellness scores, and higher physician-assigned scores. Graphically, the distribution for each of the scores overlapped as expected. In each comparison, there were a small number of “mismatches” with better or worse perceived severity than the SSS suggested. This pattern was seen with each of the paired comparisons (Figure 3).

Physical Exam Findings

Ninety-three percent of patients had a normal exam result. Joint hypermobility was noted in 1% with a mix of other less specific abnormalities in 3.4% and incomplete data forms in 13%. Pathologic murmurs were rare, noted in <1%. There was no significant difference between groups ($\chi^2 P=0.70$).

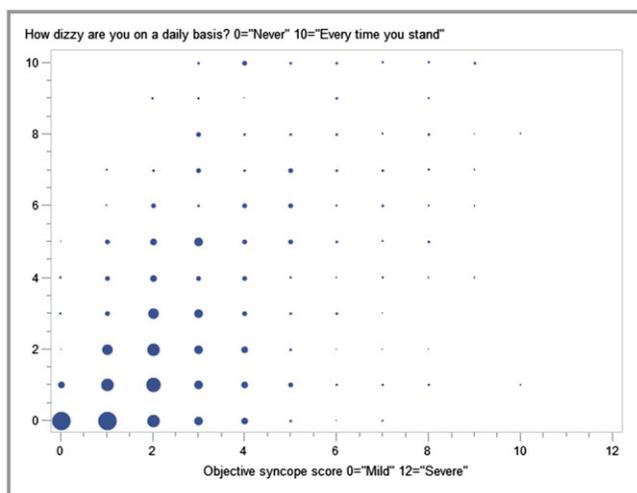


Figure 3. Weighted circle plot of correlating combinations of the 2 different symptomatic scores. The size of the circle reflects the number of responses. This example demonstrated the 0 to 10 dizziness/syncope score compared to the 12-point ad-hoc physician-assigned scores. The pattern was similar for each pairing. All had strong clustering at the “well” end of the scale. Each score had patients who had mismatches between the different scales.

Orthostatic Vital Signs

The Phase 1 pathway recommended sitting heart rate and blood pressure with formal orthostatic vital signs using the American Autonomic Society guidelines of supine values at 1 and 3 minutes followed by standing values at 1 and 3 minutes.¹⁶ Each specific value was recorded in 40% to 76% of patients with typical syncope and in 75% to 94% of patients diagnosed with POTS. For the initial SDF, a change in HR between supine and standing of ≥ 30 bpm combined with plausible symptoms permitted a diagnosis of POTS.

For the overall cohort, the baseline supine HR was 73 ± 13 with blood pressure of 111/60 (± 11 mm Hg for systolic blood pressure [SBP] and ± 9 for diastolic blood pressure [DBP]). There was no clinically meaningful change between the 2 supine measurements. There was a mean $22 (\pm 14)$ bpm increase in HR by 3 minutes of standing ($P < 0.0001$). SBP did not change with standing, while DBP increased a mean of 5 mm Hg (95% CI 3.9–5.8, $P < 0.0001$).

Supine SBP and DBP were statistically slightly lower compared to sitting (SBP 111 ± 12 ; DPB 63 ± 9 , $P < 0.01$) but the magnitude of that change was sufficiently small (95% CI 0.4–2.1 mm Hg for SBP and 2.0–3.5 for DBP) that these distinctions were judged insignificant in a practical sense. With continued standing between the first and third minutes, there were minimal changes in paired systolic BP measurements (an ≈ 2 mm Hg decline between 1 and 3 minutes) with no change in mean measurements.

A summary of orthostatic HR changes is presented in Figure 4. Overall, 79% of typical syncope patients had

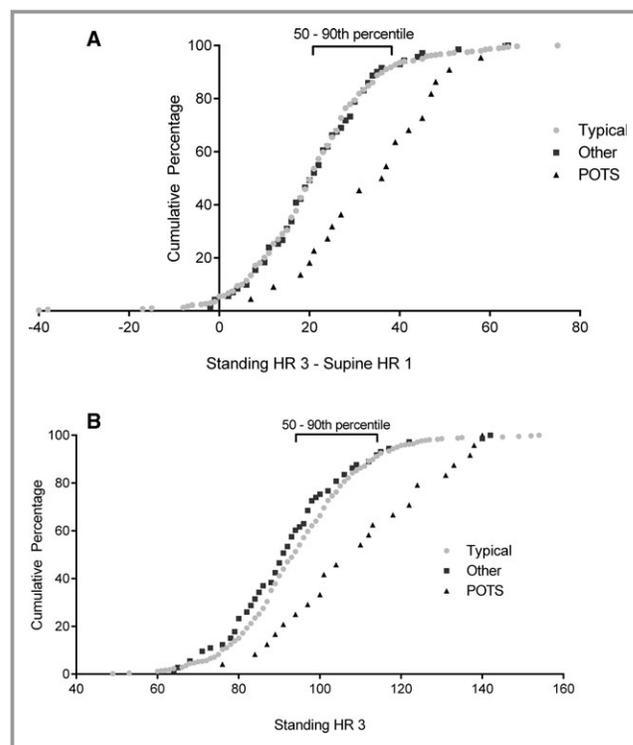


Figure 4. Cumulative histograms of absolute heart rate (A) and change in heart rate between 3 minutes of standing and the initial supine heart rate (B). For typical syncope, the 90th percentile of standing heart rate was 115 bpm with the 93rd percentile at 120. Similarly, for the change in heart rate 35 bpm represented the 90th percentile. For comparison, those classified as postural orthostatic tachycardia had a wide range, though a reasonable percentage were well within the normal changes. HR indicates heart rate; POTS, postural tachycardia syndrome.

< 30 bpm and 94% had < 40 bpm increase from supine heart rate at 1 minute to standing heart rate at 3 minutes compared to 36% of patients classified as POTS who had < 30 bpm and 64% < 40 bpm increase, respectively.

The relationship between orthostatic changes and clinical classification of POTS was examined with a series of receiver operating characteristic curves. This analysis supported using the difference between standing 3 minutes and any of the supine measures (1 minute, 3 minutes, or the minimum) as most predictive of this clinical classification, albeit with an only modest precision evidenced by an area under the receiver operating characteristic curve of 0.76 to 0.79. Receiver operating characteristic analyses for SBP and DBP showed no ability to discriminate.

ECG Testing

There was high adherence to obtaining an ECG, with 98% of those presenting reporting results. Overall 90% were normal.

ECGs were intended to be evaluated using criteria that mirror the Seattle Criteria¹⁷ included on the SDF. A detailed listing of abnormalities in typical syncope (Table 3) revealed multiple “borderline” findings. Those findings were a minor contributor to use of echocardiograms. Proportionally more ECGs were reported to be abnormal (16%, 21%) in the relatively small number of patients presenting with exertional and atypical/refractory syncope (Fisher Exact $P < 0.0001$). Borderline QTc intervals (7) and an ECG with a putative Brugada pattern were identified that would appropriately require additional evaluation in 0.6% of the ECGs. By design, those with diagnostic findings of WPW and QTc > 480 ms were excluded from the SCAMP.

Ambulatory Monitoring

One hundred and twenty-four patients underwent ambulatory monitoring. Only 62 results were reported, with 87% of these reported as normal and 8 (13%) with incidental findings. No results explained the syncope.

Echocardiogram Findings

Echocardiograms were performed in 331 patients (26%), with indications for echocardiograms summarized in Table 4. Indications were not recorded in 14%. A single patient in the exertional group was found to have cardiac syncope, with hypertrophic cardiomyopathy. A post-hoc analysis was done based on the reported findings, syncope classification, and comments. Including that analysis, 85% of the echocardiograms performed were done in compliance with the SCAMP protocol, and 5% were performed for family or primary care physician anxiety. The vast majority (226, 88%) were normal with scattered incidental findings seen in 11% (Table 5). With the single exception, none of those findings were felt to explain the syncope. They ranged from completely trivial to findings that may drive additional follow-up. Data were not collected on the clinical response to these findings.

Ferritin levels and laboratory testing

There was low compliance with this recommendation. Of the 188 patients with typical syncope scaled as moderate to severe, 71 (38%) had no lab work obtained with only 54 (29%) having both complete blood count and a ferritin level as recommended. Overall 94 (8% of total) had ferritin levels obtained. Of the 14 patients with mild typical neurally mediated syncope and ferritin lab results, 6 (43%) had ferritin < 50 ng/mL, suggesting low iron body stores. No follow-up was available after intervention with iron supplementation.

Table 3. ECG Abnormalities

Abnormality	n (%)
Total	96
LVH	17 (18)
Bundle branch block	12 (13)
RVH	11 (11)
Inverted T-waves	9 (9)
QRS axis < -20 and > 130	7 (7)
QTc 450 to 479 ms	7 (7)
RAE or LAE	3 (3)
Ventricular ectopy	3 (3)
ST-T segment change > 2 mm	2 (2)
PR interval > 220 ms	2 (2)
Atrial ectopy	2 (2)
Brugada pattern	1 (1)
Other abnormality	39 (41)
Sinus bradycardia	10
Nonspecific ST-T wave changes	7
Right axis deviation	4
Left axis deviation	3
First-degree AV block	3
Atrial premature beats	2
Borderline LVH	2
Flattened T waves—low voltage	1
Sinus arrhythmia	1
Sinus rhythm with RSR prime, not meeting RVH criteria	1
Pathologic Q waves, decreased RV side forces	1
RSR prime pattern in V1 and V4R consistent with right ventricular hypertrophy	1
Borderline RAE, want f/u ECG	1
Pure R in V1	1
RSR' c/w right ventricular volume overload	1

AV indicates atrioventricular; c/w, consistent with; f/u, follow-up; LAE, left atrial enlargement; LVH, left ventricular hypertrophy; RAE, right atrial enlargement; RVH, right ventricular hypertrophy.

Follow-Up

There were a total of 143 follow-up encounters representing 10% of the total visits. One hundred and one (71%) of these encounters were follow-up for typical syncope. Deviations in practice and patient behavior were instructive. While 127/843 (15%) of patients with a low-severity typical syncope score were recommended for follow-up, only 34 (4%) returned. Overall, cumulative follow-up rates were higher for the 379 patients with other syncope types. Largely because typical low-severity syncope is so common,

Table 4. Indications for Echocardiogram

	Patient's Description of Syncope N (%)	Abnormal Physical Exam N (%)	Abnormal ECG N (%)	Concerning Family History N (%)	Family/PMD Anxiety N (%)	Other N (%)	Not Recorded N	Total
Total	117 (35)	16 (5)	75 (23)	30 (9)	17 (5)	82 (25)	46	331
Mild typical	21 (12)	10 (6)	57 (32)	20 (11)	7 (4)	50 (28)	31	176
>Mild typical	5 (15)	2 (6)	3 (9)	3 (9)	4 (12)	10 (30)	10	33
Atypical/refractory	20 (69)	1 (3)	7 (24)	3 (10)	3 (10)	11 (38)		29
Exertional	61 (88)	2 (3)	5 (7)	1 (1)	1 (1)	4 (6)	3	69
Convulsive	9 (50)		2 (11)	2 (11)	2 (11)	5 (28)	1	18
POTS	1 (17)	1 (17)	1 (17)	1 (17)		2 (33)	1	6

PMD indicates primary medical doctor; POTS, postural tachycardia syndrome.

89 (62%) of the follow-up encounters that occurred were for SSS ≤ 4 .

Adherence to Initial Guidelines

The pathway flow charts were analyzed to create deviation reports. We focused on the typical syncope. Overall adherence was excellent, with >90% adherence for initial evaluation and most specific decision points. Notable findings included the lack of clarity in 3 of the history questions with one question eliminated from analysis.

Overall testing deviation is summarized in Table 6. For typical low-severity syncope, the most frequent deviation was the choice to obtain an echocardiogram. Only in a minority of encounters were echoes not obtained when recommended. This practice persisted for echo for other types and severity of syncope. For other cardiac testing (Exercise, Ambulatory ECG) the frequency of deviations was lower, while for lab testing the frequency of deviations was higher. There were significantly more deviations in those encounters with syncope that was not typical, low-severity syncope ($P=0.02$ for echo, $P<0.01$ for exercise test, ambulatory ECG, and lab testing). Of patients with deviations, there were significantly more deviations to obtain a nonrecommended test in typical low-severity syncope compared to other syncope classifications. In other classifications, the deviations were more frequently to defer testing ($P<0.01$ in all 3 cases). There was some variability in the use of orthostatic vital signs, with less frequent completion of those in patients with typical low-severity syncope. Additionally, as noted above, follow-up recommendations varied both in clinician and in patient acceptance. Taken together, that behavior suggested less follow-up was required.

An abnormal ECG was intended to trigger an echocardiogram; however, in 35% of those cases an echocardiogram was not ordered, further emphasizing the marginal perceived utility of echocardiography and/or of borderline ECG findings.

Discussion

During the first 2 years of a regional SCAMP implementation focusing on childhood and adolescent syncope, 85% of the patients enrolled were diagnosed with typical syncope. This cohort had the expected demographics with an average age of 14.5 years and the majority of patients female.⁴ Typical syncope was by far the most common diagnosis for both sexes. There was a trend toward a higher frequency of males with convulsive syncope and females with exertional syncope. Typical neurally mediated syncope had the expected historical features although potentially high-risk characteristics such as injury, incontinence, and syncope around exertion occurred at moderate frequency without obvious heart disease.

This study documents the largest systematic use of orthostatic vital signs for the evaluation of syncope in pediatric patients. Noteworthy is the large variability in orthostatic HR response in patients with typical syncope (20% demonstrated >30 bpm increase in HR by 3 minutes and 7% a 40 bpm increase). Our findings confirm the recent shift to requiring higher cutoff values of >40 bpm in pediatric and adolescent patients to make a diagnosis of POTS.^{16,18} An important practical new finding is the observation that orthostatic blood pressure changes between supine, sitting, and standing were clinically trivial in all syncope groups and therefore deemed not useful in this setting. We propose a streamlined workflow for initial visits that includes a single seated HR and blood pressure. For those where the possibility of POTS is being considered, use of supine and 3-minute standing HR measurements (without obtaining blood pressure) is reasonable with a cutoff of a 40 bpm increase and a standing HR of >115 to 120 at 3 minutes distinguishing a normal versus exaggerated response to orthostatic pulse in an office setting.

This SCAMP used 3 simple syncope severity scores. Patient-assigned subjective scores on dizziness and well-being, modeled after Rowe's early experience with POTS and

Table 5. Incidental Abnormalities Found by Echocardiogram

Syncope Type	Incidental Abnormality
Mild typical	Borderline MVP, no MR
	Tricuspid valve prolapse
	Mild central aortic regurgitation, mild ascending aortic dilation
	Mild aortic dilation
	Premature ventricular contractions
	Borderline apical LV noncompaction
	Coronary arises from ST junctional, normal courses (??)
	Trivial AI, normal aortic valve
	Secundum atrial septal defect
	Mild aortic regurgitation and likely partial fusion of his aortic valve commissure
	Small fenestration in the atrial septum with left-to-right flow
	Moderate TR and tiny patent foramen ovale
	Mildly dilated LV, low normal function
	Bicuspid aortic valve (mild aortic insufficiency, no aortic stenosis). No root dilation.
	Mild central aortic regurgitation and mild descending aortic dilation
	Mild LV dilation
Normal structure, but some ectopy	
Mild tricuspid regurgitation. Low right ventricular pressure.	
Hypertrabeculation pattern	
>Mild typical	Mild pulmonary branch stenosis
Exertional	High takeoff of right coronary
	Top normal to mild root dilation
	Trivial aortic regurgitation
	Possible small patent foramen ovale
	Aortic insufficiency
	Partial fusion of L & R coronary cusp
	Mildly dilated left ventricle, normal mass:volume ratio
	Small ASD
Small left coronary system of concern, but not definitive	

AI indicates aortic insufficiency; ASD, atrial septal defect; LV, left ventricular; MR, mitral insufficiency; MVP, mitral valve prolapse.

Chronic Fatigue,¹⁹ generally correlated well with an ad-hoc physician-assigned 12-point scale. The clinician behavior and patient follow-up choices supported a physician-assigned score of ≤4 representing low-severity syncope. Practically, this designation included up to 2 to 3 episodes of syncope with minimal school loss, no injury, and no late residual symptoms. This physician-assigned SSS correlated with the

self-assigned scores for well-being and dizziness. These data support using simple scores to help assess the level of disability (whether perceived or absolute) for their syncope symptoms. The SSS summarizes the actual and perceived disability related to the syncope syndrome and likely will help predict which patients may be interested in therapy or follow-up. Importantly, because of the exceptionally low frequency of identified cardiac disease, these scores do not affect probability of critical cardiac diagnoses. The 12-point SSS requires some modification to effectively reflect follow-up visits, as the number of faints was not limited to a defined time period.

There are ample data to support that typical neurally mediated syncope in the school-age child and adolescent results in excessive testing,^{20–22} with a systematic trend toward more testing in the last 40 years.⁴ Adult groups have developed reasonable predictive rules; however, they also have disease incidence and severity that are orders of magnitude higher than seen in pediatrics.²³ Given the low prior probability of significant disease, once the ECG is nearly normal, serious cardiac disorders will always be rare in pediatrics.^{24,25} Using SCAMP methodology, we can therefore decrease practice variation and focus toward both an efficient and a safe approach.

The ECG was asserted to have a high negative predictive value for serious heart disease²⁴ with early exclusion of patients with WPW and Long QT Syndrome (defined as QTc ≥480 ms). Using predefined criteria, ≈10% of the ECGs demonstrated a wide range of “borderline” abnormalities not likely contributing to syncope. An abnormal ECG was a minor contributor to the decision to perform an echocardiogram. The one new cardiac diagnosis had both high-risk symptoms and a clearly abnormal ECG with T-wave inversion and left ventricular hypertrophy. Interestingly, 16% of ECGs in patients with exertional symptoms were read as abnormal. These may be systematic changes in athletes’ ECGs, but could also suggest that cardiologists may interpret the ECG differently when evaluating a patient with a potentially higher probability of significant disease. Neither bias would change practice, as history drives the decision to obtain an echocardiogram or other testing in those with exertional syncope.

The exclusion of WPW and LQTS, 2 of the most obvious ECG diagnoses, represents the reality that in clinical evaluation of a patient, any of those findings will drive decision making away from typical syncope. A small number (7 borderline QTc and 1 potential Brugada pattern or 0.6%) of the ECGs suggested additional investigations for ion channel disorders. The overall prevalence of those disorders (≈1/700 for WPW and ≈1/2000 for LQTS) informs choices about whether an ECG is “mandatory” following an event that otherwise meets clinical criteria for typical, low-severity, neurocardiogenic syncope.

Table 6. Deviation Table

Test	Total With Known Recommendation	% Deviated From Recommendation	% With Test Against Recommendation	% Without Test Despite Recommendation
Overall				
Echo	1228	20	16	4
Exercise test	1209	10	3	7
Ambulatory ECG	1214	9	6	4
Ferritin Lab	1254	13	2	11
Mild typical				
Echo	861	18	14	4
Exercise test	875	2	2	0
Ambulatory ECG	875	5	5	0
Ferritin Lab	875	2	2	0
All others				
Echo	367	23	20	3
Exercise test	334	30	4	26
Ambulatory ECG	339	21	8	13
Ferritin Lab	379	41	3	38

The vast majority (85%) of echocardiographic studies were performed for historical details or other clear SCAMP indications. The most common indication for echo was the description of the syncopal event in 35% of patients. Echo was performed less frequently in those with typical syncope, only 20% compared to 64% of those with other than typical syncope. Given the larger volume of patients with typical syncope, these represented 63% of the echoes performed. Similar to the chest pain SCAMP,⁷ incidental echocardiogram findings that did not explain the syncope were identified in 11%.⁷ The high frequency of deviations from the proposed guidelines emphasizes the variations in perceived marginal utility of echo. The need for a post-hoc analysis to review echo indications demonstrated limitations in both the SDF and likely the nature of decision making. Interestingly, during the chest pain SCAMP \approx 40% of patients had echocardiograms.⁷ While echo use did not decline over the period of use of the syncope SCAMP, the overall use of echocardiography was nearly half that of the chest pain SCAMP, which immediately preceded this effort and included substantial overlap among practitioners. Even in the peri-exertional syncope group, there was some deviation away from doing an echocardiogram. While speculative, it is plausible that the experience and comfort with deferring an echocardiogram in the chest pain cohort contributed to this behavior. Overall this may be part of increased recognition that all testing has the potential to identify incidental findings,²⁶ which may increase anxiety or even morbidity.

There remains substantial practice variation in the use of exercise testing and ambulatory ECG monitoring. Both have significant limitations as diagnostic tests and carry with them important logistical concerns that increase the burden on families/physicians. Given the low yield of either in identifying serious disease, future efforts will aim at much more selective use of these studies.

While the follow-up volume and duration was somewhat limited, these data showed that fewer people were recommended to return than suggested on the initial guideline and that fewer still actually returned for follow-up. This behavior, quite likely, represents the appropriate follow-up and suggests that a reassuring “come back if you have more problems” approach is fully justified.

The low incidence of identified cardiac disease, combined with Kane’s retrospective analysis demonstrating the effectiveness of identifying life-threatening cardiac disease using the chest pain clinical pathway,¹⁰ emphasize that clinicians are quite effective at identifying those disorders. Hence, when patients return with problematic symptoms, and without a specific feature of the history suggesting serious disease, the focus may be more on effective management of the symptoms. Fortunately, as the vast majority of patients do not need to return, even this large experience cannot provide useful data on more advanced and pharmacologic management.

Two related observations raise the question of whether patients with high syncope scores and more disability (atypical, POTS, and high-severity typical syncope) represent

a comprehensive pediatric “problem” more than a subspecialty cardiology problem. These subgroups had high use of prior specialty referrals and a high use of medications. There are at least 3 potential causes for that association, which cannot be answered here. These patients may have a medical or psychiatric diagnosis that makes them more prone to syncope; their therapy may predispose them to syncope (certainly a common cause of syncope in the elderly), and lastly, they may be excessively medicalized with syncope part of a pattern of somatization. All of those potential causes suggest careful evaluation of problematic patients in their medical home prior to or coincident with referral to cardiology.

The Boston Children’s Hospital emergency department, in parallel with these efforts, demonstrated that a systematic approach to syncope patients can decrease resource utilization and limit low-yield testing.²⁷ A multicenter experience in China included a relatively large number with cardiac syncope.²⁸ With a tiered protocol that included frequent head-up tilt testing, typical syncope was identified in 73% of patients. They assert that this approach decreased resource utilization, even if they were more interventional than current local practice. The guideline used here specifically accepts that typical low-to-moderate-severity syncope is almost certainly neurocardiogenic, and does not require confirmatory testing. That choice was made both because head-up tilt testing is increasingly discredited as a routine test for typical syncope²⁹ and to limit the medical burden associated with physiology that overlaps with normal.³⁰ This workflow permits some uncertainty regarding the precise noncardiac diagnosis, with the trade-off of limiting patient, family, and medical resource use. Implicit in that overarching plan is that more complicated pictures (whether POTS, somatoform, or difficult-to-identify arrhythmias) will have recurrent symptoms and can return for care.

Limitations

Serious cardiac disorders both are likely identified immediately and may present less frequently in the office setting. By design, WPW and Long QT Syndrome, the 2 diagnoses most apparent on ECG, were excluded from the SCAMP. Clinicians chose to enroll patients in this process, so that if they immediately classified them as having heart disease the patients are not included in this experience. This initial report focuses on the majority with typical syncope and cannot provide significant data on the more problematic patients with either other forms of syncope or those who had follow-up recommended or chose to return for follow-up.

Explicit in an approach like this is that there will always be rare diseases that either are not seen on ECG or exam (anomalous coronary, catecholaminergic polymorphic VT as 2

classic examples). Both are rare, both are classically associated with at least peri-exertional if not midexertional symptoms,^{31,32} and hence, in this clinical pathway, should be detected with proscribed testing. All diagnostic testing carries with it the risk of incidental findings, false positive/false negative results, and ineffective interpretation (whether a technical or provider issue). While these data incompletely tabulate the burden of “positive” tests, they do not address those issues. Test results are not adjudicated or confirmed. Rather, the SCAMPS approach focuses on the behavior of the clinician in reacting to the data.

During pathway development, we invested significant energy in suggesting more advanced therapy options and somewhat creative approaches to advanced testing. The paucity of patients with more persistent symptoms limited further analysis. The limited data on ferritin levels support the idea that considering deficient iron stores may be a contributor to more symptomatic patients.^{13,14} The poor adherence to those recommendations similarly suggested (at least to the cardiologists) that iron studies are best done in a primary care setting. The enthusiasm for developing these more advanced options speaks to the challenge of treating patients with higher SSS despite representing a small group.

Experienced clinicians rapidly identify precise details of the history that shift their thinking toward more serious diagnoses.^{5,32,33} Given the low incidence of serious disease, this experience has a limited role in refining that algorithm. The frequency of palpitations, injury, and incontinence seen only emphasizes that those findings have a limited positive predictive value.

Conclusions

Using SCAMP methodology, we have demonstrated that typical neurally mediated syncope represents the vast majority of children referred to pediatric cardiologists for syncope. Cardiac etiology is rare, and in this study one patient with hypertrophic cardiomyopathy was identified using the suggested evaluation plan. Syncope can be evaluated in the ambulatory setting pragmatically and effectively using minimal resources and in the vast majority in a single initial visit.

This experience confirms and solidifies many of the traditional historical details that define typical neurally mediated syncope. Subjective and physician-assigned Syncope Severity Scores appear to provide a rapid way to stratify patients likely to require additional care. The relatively high frequency of potentially concerning family history, mild abnormalities on ECG, and incidental findings on echocardiography and ambulatory monitoring serve as a cautionary note regarding the need to place both the decision to obtain a test, and its interpretation in context.

Finally, our findings demonstrate that orthostatic vital signs add little if anything to the initial evaluation. Those should be reserved for more symptomatic patients in whom a streamlined workflow with a single baseline blood pressure followed by use of supine and 3-minute standing HR measurements is appropriate. Using that technique, a standing HR <120 bpm and a change from supine to standing of <40 bpm is within the 93rd percentile, and <115 bpm and HR change of <35 bpm is within the 90th percentile. Those ranges provide data-driven and easy-to-remember thresholds for those in whom orthostatic intolerance is the more likely contributor to daily symptoms.

Evolving and explicit clinical pathways such as SCAMPS can be implemented regionally across a wide range of clinical practice settings, and this approach can overcome a number of barriers often limiting traditional clinical guideline implementation and prospective randomized controlled trials. They represent a way to relatively rapidly gain and disseminate experience among providers and centers.

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Disclosures

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