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CLINICAL RESEARCH SHORT REPORT

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Home-based multi-sensory and proximal strengthening program to improve balance in Charcot–Marie–Tooth disease Type 1A: A proof of concept study

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Abstract

Introduction/Aims: People with Charcot–Marie–Tooth Disease (CMT) frequently report problems with balance, which lead to an increased risk of falls. Evidence is emerging of training interventions to improve balance for people with CMT, but to date all have relied on clinic-based treatment and equipment. This proof-of-concept study explored whether a multi-modal program of proprioceptive rehabilitation and strength training can be delivered at home, to improve balance performance in people with CMT Type 1A.

Methods: Fourteen participants with CMT Type 1A were recruited into this randomized, two-arm study. Baseline assessments included measures of disease severity, posturography, physical function, and patient-reported outcome measurements. All participants received one falls education session. Participants were randomized to either 12 weeks of balance training or 12 weeks of usual activities. The intervention comprised a home-based, multi-sensory balance training and proximal strengthening program, supported by three home visits from a physiotherapist.

Results: Thirteen participants completed the study. The intervention was successfully implemented and well tolerated, with high participation levels. Functional measures of balance and walking showed strong effect sizes in favor of the training group. Posturography testing demonstrated moderate improvements in postural stability favoring the intervention group. Inconsistent changes were seen in lower limb strength measures.

Abbreviations: 10MTW, 10-m walk test; C7 PL, body sway path length; C7 VEL, body sway velocity; CMT, Charcot–Marie–Tooth disease; CMTES, Charcot–Marie–Tooth Examination Score; COP PL, center of pressure path length; COP VEL, center of pressure velocity; FGA, Functional gait assessment; HADS, Hospital anxiety and depression scale; IPAQ, International Physical Activity Questionnaire; QoL, Quality of life; SF36, Short Form 36.

Discussion: The intervention was feasible to implement and safe, with some evidence of improvement in balance performance. This supports future studies to expand this intervention to larger trials of pragmatic, home-delivered programs through current community rehabilitation services and supported self-management pathways.

KEYWORDS
Balance, Neurorehabilitation, Charcot-Marie-Tooth Disease

1 | INTRODUCTION

Poor balance and postural instability are often observed in people with Charcot–Marie–Tooth disease (CMT) due to muscle weakness, sensory impairment, and foot deformity.[1–6] Two studies have reported falls events in CMT cohorts of 50%[1] and 86%.[4]

Effective dynamic and static balance requires a complex interplay between motor control and the somatosensory, vestibular, and visual systems.[7] Proprioceptive impairment and distal leg weakness have been correlated with static balance impairment for people with CMT Type 1A.[5,6] Proximal disuse atrophy has also been observed,[8] with knee extensor strength being linked to forward reach distance.[9]

People with CMT are able to strengthen proximal lower limb muscles[10–13] and dynamic balance training interventions have demonstrated positive effects on balance performance in CMT and sensory neuropathy.[14–17] These studies use clinic-based training methods, some with specialized equipment, and we suggest that training people at home will reduce burden of travel, promote self-management, and be better incorporated into daily lifestyles.

In this proof-of-concept study, we propose that a rehabilitation program of multi-sensory balance training and proximal strengthening exercises can be feasibly delivered in a person's own home.

2 | METHODS

A single-blinded randomized, two-arm design was used in this Phase 1 study. Ethical approval was granted (NRES Research Committee REC reference number 16/LO/0720) and informed consent was acquired from all participants.

2.1 | Study population

Adults were recruited from a specialist, neurological center. This study focused on people with CMT Type 1A to reduce group variability in this small cohort. They were included if they had a genetic diagnosis of CMT Type 1A, a history of falls and could walk for 50 m, with aids if needed. Participants were excluded if there were co-morbidities that effect balance and walking.

2.2 | Intervention

2.2.1 | Allocation

A single, one-to-one falls education session was delivered to all participants, who were then randomized to the control or intervention group, on completion of the baseline measurements (Figure S1). The control group were advised to continue with usual activities. The intervention group received a bespoke, 12-week exercise prescription of home-based multi-sensory balance rehabilitation and proximal strengthening exercises. An unblinded physiotherapist (L. E. L.) performed a risk assessment in the participant's home and delivered the program through three home visits and weekly phone calls.

2.2.2 | Multi-sensory balance training

Training was designed to address known contributors to balance impairment for people with CMT (Table S1). Postural stability was challenged daily using sensory and mechanical perturbations to train proactive and reactive balance responses. The program was individualized to baseline ability by altering sensory feedback and base of support and progressed in difficulty using these principles (Table S2).

2.2.3 | Strength training

Individualized, proximal lower limb and trunk, body weight-based, resistance exercises were performed on alternate days. Progression was achieved through increasing the difficulty of the anti-gravity position, degree of body weight, and time under tension. Exercise selection was a collaborative process and tailored to participant's preferences (Table S3).

2.2.4 | Self-management

The intervention was underpinned by an established self-management support framework and focused on augmenting engagement in the home-based training by promoting self-management techniques such as collaboration, problem solving, reflection and self-discovery.[18]

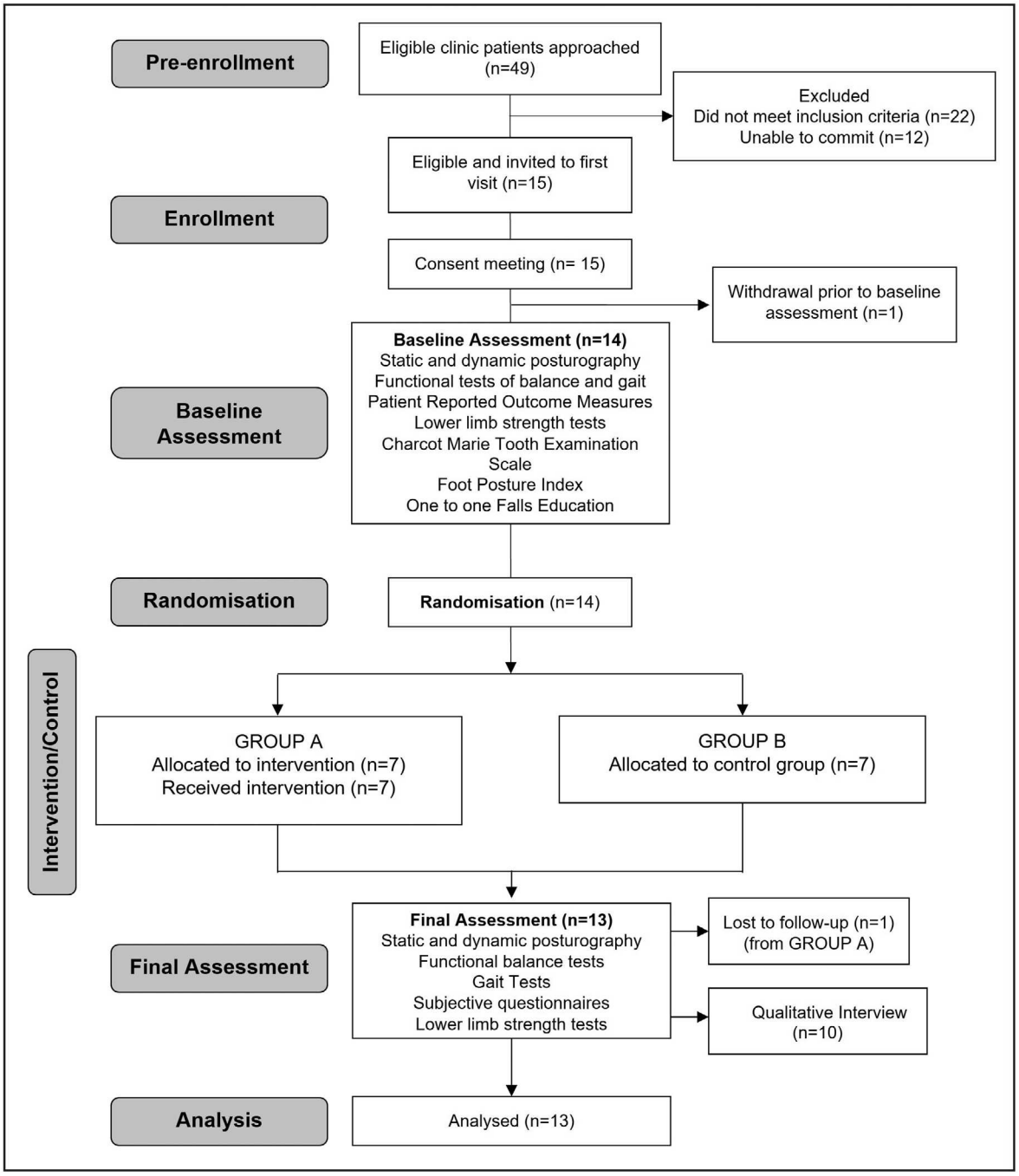

FIGURE 1 Consort flow diagram.

Participants were supported to identify safe and effective ways to integrate the intervention into their daily routines and set personalized goals around engagement.

2.2.5 | Monitoring

The intervention group received 12 weekly phone calls and the control group 3 monthly calls.

2.3 | Outcomes

2.3.1 | Feasibility and safety

Engagement with the program was monitored using weekly phone calls asking set questions to record program participation rate, challenges, and any adverse incidents (Table S4). This was supported by a smartphone app. Participants were also invited to individual, semi-structured interviews after the study (Table S5).

TABLE 1 Participant baseline demographics.

Baseline demographics	Group A intervention ($n = 7$)	Group B control ($n = 7$)	Significance (Mann–Whitney *U* test)	
			p-value	*z*-score
Age (years)	44 (30–65)	53 (36–70)	0.18	1.34
Sex			0.70	0.38
Female	5	6		
Male	2	1		
Height (m)	1.7 (1.62–1.79)	1.7 (1.59–1.73)	0.28	1.09
Weight (kg)	85.2 (53–108)	72.3 (58–91.3)	0.37	0.89
CMTES	13 (8–17)	11.6 (6–18)	0.52	0.64

Abbreviation: CMTES, CMT Exam Score.

2.3.2 | Demographics

At baseline, age and sex of participants was recorded with disease severity measured using the CMT Exam Score (CMTES).[19]

2.3.3 | Balance and gait performance

Baseline measurements were conducted by blinded evaluators (M. M. D., C. M.) after enrolment and after the 12-week intervention or control period (Figure S1).

Functional balance and mobility were assessed using the Berg Balance Scale[20] the BEST Test,[21] the Functional Gait Assessment (FGA),[22] and the 10-m walk test (10MWT) at self-selected and fast pace.[23]

Postural stability was measured using static posturography (details in Table S6) with the following variables recorded over 30-s standing trials: center of pressure path length (COP PL); center of pressure velocity (COP VEL); body sway path length (C7 PL); body sway velocity (C7 VEL).[24]

2.3.4 | Muscle function

Lower limb strength was assessed with hand-held dynamometry using a "make-test" protocol (CITEC Handheld Dynometer model CT 3002, Netherlands).[25]

2.3.5 | Patient-recorded outcome measures

Participants completed the Walk-12 scale measure perceived walking ability,[26] the Short Form 36 (SF36)[27] for health-related Quality of Life (QoL), International Physical Activity Questionnaire (IPAQ),[28] Hospital Anxiety & Depression Scale (HADS),[29] and the Falls Self efficacy scale[30] to explore fear of falling.

2.4 | Analysis

Feasibility was explored through participation in the program, as a proportion of the total prescribed sessions completed. Post-intervention, individual interviews were transcribed, and qualitative data were analyzed using thematic analysis.[31]

The effect of the intervention on quantitative measures was examined using the Hedges G effect size calculation due to the small sample. Effect sizes were categorized as strong if 0.7 or over, moderate if 0.3–0.69, and small if 0.29 or less.

3 | RESULTS

3.1 | Participant recruitment and engagement

Fifteen participants were recruited to this study, with two dropouts (Figure 1). There were no significant differences in demographics and disease severity (CMTES score) between the intervention and control groups (Table 1).

3.2 | Feasibility and safety

There was high engagement with 91% mean completion of strength exercises and 79% of balance exercises, and there were no adverse incidents. Interviews highlighted several themes relating to the intervention: Participants appreciated the individualized nature of the exercise prescription and acknowledged benefits of doing them at home. The physiotherapist was a key motivating and reassuring factor. Some participants described positive learning about their balance impairments, increasing their confidence in managing risk. Conversely, other participants described feeling more vulnerable and aware of their unsteadiness.

TABLE 2 Mean differences pre and post intervention or control, with Hedges G effect size statistic.

		Intervention			Control			
Category	Variable	Week 0	Week 12	Difference	Week 0	Week 12	Difference	Hedge's G
Posturography: Feet apart, eyes open	COP path length (m)	0.54 ± 0.42	0.36 ± 0.33	−0.18 ± 0.36	0.33 ± 0.24	0.20 ± 0.10	−0.13 ± 0.15	0.23
	C7 path length (m)	0.40 ± 0.36	0.23 ± 0.22	−0.17 ± 0.27	0.25 ± 0.22	0.24 ± 0.24	−0.01 ± 0.32	0.53[a]
	COP velocity (m/s)	2.28 ± 1.79	1.36 ± 1.24	−0.88 ± 1.14	1.24 ± 0.90	0.77 ± 0.38	−0.48 ± 0.57	0.48
	C7 velocity (m/s)	1.69 ± 1.54	0.88 ± 0.83	−0.81 ± 1	0.95 ± 0.81	0.90 ± 0.93	−0.05 ± 1.2	0.68[a]
Posturography: Feet apart, eyes closed	COP path length (m)	0.78 ± 0.49	0.56 ± 0.48	−0.22 ± 0.33	0.40 ± 0.33	0.32 ± 0.20	−0.08 ± 0.16	0.58[a]
	C7 path length (m)	0.51 ± 0.40	0.36 ± 0.38	−0.15 ± 0.27	0.30 ± 0.31	0.20 ± 0.10	−0.10 ± 0.23	0.24
	COP velocity (m/s)	3.16 ± 2.10	2.17 ± 1.83	−0.99 ± 1.18	1.50 ± 1.26	1.21 ± 0.74	−0.29 ± 0.61	0.80[a]
	C7 velocity (m/s)	2.06 ± 1.65	1.38 ± 1.43	−0.67 ± 1.01	1.12 ± 1.16	0.76 ± 0.37	−0.36 ± 0.85	0.34
Posturography: Feet together, eyes open	COP path length (m)	0.61 ± 0.35	0.44 ± 0.34	−0.17 ± 0.27	0.42 ± 0.41	0.25 ± 0.10	−0.18 ± 0.33	0.02
	C7 path length (m)	0.42 ± 0.31	0.35 ± 0.34	−0.07 ± 0.34	0.42 ± 0.68	0.19 ± 0.68	−0.23 ± 0.61	0.02
	COP velocity (m/s)	2.30 ± 1.30	1.70 ± 1.32	−0.6 ± 0.97	1.60 ± 1.57	0.94 ± 0.37	−0.66 ± 1.26	0.05
	C7 velocity (m/s)	1.57 ± 1.18	1.34 ± 1.30	−0.23 ± 1.21	1.59 ± 2.62	0.72 ± 0.48	−0.87 ± 2.38	0.32
Posturography: Feet together, eyes closed	COP path length (m)	1.04 ± 0.62	0.79 ± 0.52	−0.25 ± 0.36	0.71 ± 0.46	0.47 ± 0.21	−0.25 ± 0.26	0.01
	C7 path length (m)	0.73 ± 0.63	0.51 ± 0.45	−0.21 ± 0.39	0.52 ± 0.42	0.27 ± 0.12	−0.25 ± 0.32	0.1
	COP velocity (m/s)	3.93 ± 2.37	2.96 ± 1.96	−0.96 ± 1.39	2.69 ± 1.71	1.76 ± 0.80	−0.93 ± 0.99	0.03
	C7 velocity (m/s)	2.76 ± 2.41	1.93 ± 1.68	−0.82 ± 1.51	1.95 ± 1.58	1.01 ± 0.46	−0.94 ± 1.19	0.09
Isometric muscle strength (*N*)	Hip flexion	153.71 ± 76.72	157.57 ± 47.48	3.86 ± 64.82	137.36 ± 44.59	153.21 ± 42.18	15.86 ± 36.83	0.23
	Hip extension	150.93 ± 56.45	174.21 ± 62.56	23.29 ± 28.73	142.43 ± 56.61	152.79 ± 72.71	10.36 ± 33.75	0.41
	Knee flexion	89.79 ± 44.97	97.21 ± 31.45	7.43 ± 15.16	67.71 ± 25.89	87.79 ± 26.741	20.07 ± 29.13	0.54
	Knee extension	204.43 ± 85.17	202.93 ± 60.92	−1.5 ± 36.19	152.71 ± 63.90	166.14 ± 54.01	13.43 ± 46.37	0.35
	Dorsiflexion	45.43 ± 27,017	87.93 ± 80.89	42.5 ± 68.92	51.71 ± 43.99	68.64 ± 68.51	16.93 ± 27.68	0.49
	Plantarflexion	211.64 ± 133.41	193.28 ± 149.57	−18.36 ± 58.74	144.00 ± 124.52	190.00 ± 152.91	46 ± 79.84	0.92
Functional balance and walking	FGA	17.17 ± 8.82	20.42 ± 7.85	3.29 ± 4.5	18.86 ± 7.52	19.29 ± 7.43	0.43 ± 3.15	0.74[a]
	BERG	43.00 ± 16.72	47.43 ± 10.92	4.43 ± 6.73	48.71 ± 6.34	47.86 ± 6.15	−0.86 ± 3.08	1.01[a]
	BESTest	70.71 ± 33.92	11.43 ± 24.08	6.71 ± 10.84	78.14 ± 16.57	76.71 ± 20.09	−1.43 ± 5.74	0.94[a]
	10MTW Norm	13.11 ± 6.44	12.05 ± 5.62	−1.06 ± 1.33	10.45 ± 2.56	11.26 ± 3.88	0.84 ± 2.04	1.1[a]
	10MTW Max	10.49 ± 5.36	9.52 ± 4.53	−0.97 ± 1.59	8.07 ± 2.56	8.64 ± 3.20	0.58 ± 1.15	1.12[a]
Patient Reported Outcome Measures	MFES	81.57 ± 23.08	90.71 ± 23.55	9.14 ± 19.96	77.00 ± 32.54	91.43 ± 37.96	14.43 ± 25.62	0.23
	Walk-12	47.00 ± 10.92	43.26 ± 9.41	−3.71 ± 8.44	41.14 ± 8.29	40.57 ± 11.14	−0.57 ± 3.36	0.49
	SF36	101.14 ± 6.52	94.14 ± 484	−7.00 ± 6.00	100.00 ± 5.74	96.00 ± 5.29	4.00 ± 4.55	0.56
	IPAQ	1665 ± 1089	1971 ± 11.0	141 ± 1090	2582 ± 1559	1279 ± 1567	−1302 ± 2155	0.82[a]
	HADS	14.29 ± 6.29	13.00 ± 7.43	−3.14 ± 5.98	18.00 ± 7.55	15.71 ± 6.70	−2.29 ± 3.95	0.17

Note: Data expressed as mean ± standard deviation.
Abbreviations: 10MTW Max, 10-m walk at maximum speed (s); 10MTW Norm, 10-m walk at self-selected speed (s); BERG, Berg Balance Scale; BESTest, Best Test; C7 path length, C7 marker path length (m); C7 velocity, C7 marker velocity (m/s); COP path length, center of pressure path length (m); COP velocity, center of pressure velocity (m/s); FGA, Functional Gait Assessment; HADS: Hospital Anxiety and Depression Scale (total score); IPAQ, International Physical Activity Questionnaire (MET-minutes per week); MFES, Modified Falls Self-Efficacy Scale; SF36, Short Form 36.
[a]Effect sizes >0.5 (moderate to large) in favor of the intervention.

3.3 | Response to training

3.3.1 | Balance performance (functional measures)

Large effect sizes were observed in the FGA, the Berg Balance Test, and the Best Test in favor of balance training (Table 2). Large effects were also observed in 10-m walk times, with improvements in gait speed in the intervention group at a self-selected speed.

3.3.2 | Balance performance (posturography)

There were variable responses to training observed with an effect of training for the less challenging conditions. Conditions with feet apart and eyes open, showed moderate effect sizes in COP VEL, C7 PL, and C7 VEL, favoring the intervention group (Table 2). Closing eyes with feet apart showed moderate effect sizes in COP PL and COP VEL (Table 2).

3.3.3 | Muscle function

Changes in lower limb strength were inconsistent, with moderate effect sizes favoring the intervention group for hip extension and ankle dorsiflexion. Moderate effect sizes favored the control group for knee flexion and knee extension with a large effect for plantarflexion (Table 2).

3.3.4 | Patient-reported outcome measures

Perceived walking ability (Walk-12) and health related QoL (SF-36) demonstrating moderate effect sizes in favor of the intervention group (Table 2).

4 | DISCUSSION

In this small cohort of adults with CMT, a home-based balance intervention was feasible and safe, with some early signal for improvement in balance performance that is in keeping with clinic-based balance interventions.[14,16,17] This individualized, flexible program was well received with high engagement.

Large effects in favor of the intervention were observed in multiple functional measures, indicating a consistent finding that is applicable to daily life balance challenges[32,33] and consistent with other balance studies.[14,16] Moderate effect sizes indicated improvements in the posturography variables for the intervention group consistently in the eyes open, feet apart conditions. Balance training alone may not be sufficient to improve stability with more challenging conditions, for example, feet together and eyes closed, due to the degree of sensorimotor impairment, and we may need to look towards alternative strategies, for example, orthotics.[34,35]

There were inconsistent effects on muscle strength that may be a function of the small sample, with previous studies of strength training including samples of 18–60.[13,36] The sample sizes are insufficient at this stage to account for inter-subject variability at baseline and with training response. The large standard deviations highlight reliability issues with hand-held myometry, despite a good interrater reliability with isokinetic dynamometry.[37] MRI could also be a valuable tool to explore changes within the muscle in response to training, as used in a study of distal muscle training in children and young adults with CMT1A.[36]

5 | CONCLUSION

This proof-of-concept study demonstrated a feasible and safe alternative to hospital-based balance training, with early evidence of improvements in functional balance and gait. A larger study is required to ascertain if these positive trends translate to intervention efficacy, and we plan to develop training materials for physiotherapists to deliver the program in the community.

AUTHOR CONTRIBUTIONS

Magdalena M. Dudziec: Investigation; writing – original draft; methodology; data curation; formal analysis; software; project administration; validation; visualization. **Laurence E. Lee:** Investigation; methodology; writing – review and editing; project administration; data curation. **Charlotte Massey:** Investigation. **David Tropman:** Methodology; conceptualization; software; formal analysis; validation. **Mariola Skorupinska:** Investigation; project administration. **Matilde Laurá:** Resources. **Mary M. Reilly:** Supervision; resources; writing – review and editing. **Gita M. Ramdharry:** Conceptualization; investigation; funding acquisition; writing – original draft; methodology; supervision; resources; visualization; validation; software; formal analysis; project administration.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflicts of interest.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

ETHICS STATEMENT

We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

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SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

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