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Impact of a Pilates intervention on physical function in children with generalised joint hypermobility and chronic musculoskeletal pain: A single-case experimental design

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ABSTRACT

Aim: To assess the impact of a Physiotherapist-led Pilates Intervention for school aged children with Generalised Joint Hypermobility (GJH) on pain, physical function and Health Related Quality of Life (HRQoL).
Methods: Three children aged 8–12 years with GJH participated in an 8 week Physiotherapist-led Pilates Intervention within a single-case experimental design (multiple baseline design). Repeated measures were collected during baseline, intervention, withdrawal and follow-up, for: (i) pain, (ii) physical function as measured by muscle strength, postural control, fatigue and activity levels and (iii) HRQoL.
Results: Within the intervention phase, two children showed reduced fatigue and one child improved in muscle group strength of hip abduction (gluteus medius) and scapula adduction/rotation (rhomboideus major/minor) and HRQoL. No improvements were seen in pain or postural control. Within the early withdrawal phase all children showed improved strength for at least two muscle groups and one child showed reduced fatigue, pain (worst in last week) and improved postural control (functional reach lateral).
Conclusions: Pilates may provide an effective intervention for children with GJH to reduce fatigue and improve muscle strength and HRQoL. Limited conclusions can be made regarding pain and postural control. Further research with a longer Pilates duration is needed to confirm the dose and benefits for this population.

1. Background

Generalised Joint Hypermobility (GJH) is present when individuals have multiple joints affected by hypermobility with a distribution typically involving all four limbs and the trunk (Castori et al., 2017). The 9-item Beighton Score (Beighton, 1973) is used to classify GJH and the prevalence of GJH varies depending on the joint threshold used (Smits-Engelsman et al., 2011). For example, in an Australian study of 14 year old children, 48% of children were deemed to have GJH using the cut-off threshold of ≥4/9 hypermobile joints, whereas 18.6% had GJH with a more rigorous cut-off of ≥6/9 hypermobile joints (Morris et al., 2017). Recent literature had suggested that using a cut-off of ≥6/9 hypermobile joints may be more valid and clinically useful when working with children (Juul-Kristensen et al., 2017).

Children with hypermobility may present with a number of structural and functional problems including musculoskeletal pain (Engelbert et al., 2017; Tinkle and Levy, 2019), joint instability (Pacey et al., 2014), poorer proprioceptive acuity (Fatoye et al., 2009; Pacey et al., 2014; Smith et al., 2013), reduced muscle strength (Pacey et al., 2013; Peterson et al., 2018; Simmonds and Keer, 2007) and fatigue (M. C. Scheper et al., 2017). If these symptoms aren't managed early and effectively, it can lead to chronic disability and can significantly reduce the child's activity and participation at home, school and in the community (M. Scheper et al., 2017).

Pain is often the initial reason that a child with GJH presents to a health professional (Cattalini et al., 2015). Pain is thought to be caused through a combination of macrotrauma and microtrauma to the hypermobile joint (Castori et al., 2017). Macrotrauma includes dislocation, subluxations and any soft tissue injury. Microtrauma is caused by subtle repetitive joint injury not perceived by the individual, most likely caused by the excessive joint movement and joint instability. Pain has significant effects on a child with GJH and by 8 years of age, a strong correlation has been observed between increased pain intensity and a decreased quality of life across domains of physical, emotional, social

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and school functioning (Fatoye et al., 2012).

Fatigue is often present in children with hypermobility and can be a combination of physical fatigue due to joint pain, deconditioning and poorer muscular endurance, or mental fatigue, which is regularly associated with factors such as pain, poorer sleep and dysautonomia (M. C. Scheper et al., 2017; Tinkle and Levy, 2019). Fatigue along with pain can become quite disabling, with higher fatigue being a strong predictor of lower quality of life in this population (Mu et al., 2019; Pacey et al., 2015).

Reduction in pain is one of the major goals for children with GJH and chronic musculoskeletal pain. Management can encompass a spectrum of pharmacological, psychological and physical treatments. The focus of this study is exercise and physiotherapists with their underlying knowledge of joints, muscles and movement are well placed to direct physical interventions for the effective management of children with GJH. The current evidence base on effective exercise type, for children is limited, with one study by Kemp et al., reporting that children with GJH and pain experienced a significant reduction in pain with participation in either a 6 week generalised or targeted physiotherapy program (Kemp et al., 2010). The second study by Pacey et al., demonstrated that a Physiotherapist–led, individualised exercise program was significantly effective in reducing pain in a single joint whilst increasing muscle strength and improving HRQoL in children with GJH and chronic knee pain (Pacey et al., 2013). In a study of adults in the UK with GJH and chronic pain, 77% agreed that exercise was important for their management of pain, with swimming, walking and Pilates reported to be the most helpful modes of exercise (Simmonds et al., 2019). This finding supported investigation into the effectiveness of Pilates with children with GJH.

Our recent systematic review on the effects of a Pilates intervention in children (Hornsby and Johnston, 2020) revealed no specific literature pertaining to children with hypermobility. It did show that Pilates is effective for children in other paediatric populations with musculoskeletal pain in: reducing pain levels (Alves de Araújo et al., 2011; Mendonça et al., 2013); and improving muscle strength (Mendonça et al., 2013), postural orientation and balance (Walowska et al., 2018), musculoskeletal alignment (Alves de Araújo et al., 2011), flexibility (González-Gálvez et al., 2015; Tunar et al., 2012), physical function (Finatto et al., 2018; Tunar et al., 2012) and health related quality of life (HRQoL) (Mendonça et al., 2013). Pilates is a series of exercises developed by Joseph Pilates in the 1920's and detail regarding the history and clinical application of Pilates is illustrated in the protocol paper for this study (Hornsby, 2021). It was postulated that a Physiotherapist-led Pilates intervention using the Pilates integrated approach of treating multiple body regions simultaneously, would benefit children with GJH and chronic musculoskeletal pain by aiming to improve strength, postural alignment and motor control to effectively distribute movement load and subsequently reduce pain through involved joints. A *Single Case Experimental Design (SCED)* was used, as this study design is suitable for smaller cohorts that require intensive assessment and intervention by qualified and experienced practitioners.

The aim of this study is to research the impact of a Physiotherapist-led Pilates intervention for school aged children with GJH and chronic musculoskeletal pain on pain, physical function, activity levels and HRQoL. It was hypothesized that following Pilates, children will demonstrate a decrease in pain and fatigue with an increase in their muscle strength, postural stability, activity and HRQoL.

2. Methods

2.1. Study design and ethics

As outlined in the protocol paper by Hornsby in 2021, this study involved a *Single Case Experimental Design (SCED)*. A full description is presented in the procedure. The trial was registered with the Australian New Zealand Clinical Trials Registry (ID ACTRN12618000836235) and approved by the Human Research Ethics Committees of the Children's Health Queensland Hospital and Health Service (NHMRC EC00175) and The University of Queensland (NHMRC EC00456).

2.2. Participants

Eligible participants were children: (i) aged between 8 < 18 years; (ii) with GJH as demonstrated by ≥ 6 joints on the Beighton Score; (iii) with chronic musculoskeletal pain experienced at least 3 times per week, for a 3 month period in the previous 6 months; (iv) with pain experienced in ≥2 joints. Children were excluded if they reported a history of: (i) a diagnosis of any other heritable disorder of connective tissue; (ii) a medical or neurological condition; (iii) an intellectual impairment or behavioral disorder that may confound results; or (iv) if they had participated in a Pilates intervention in the previous 6 months. Children were recruited in a metropolitan city through fliers and expression of interest letters to public hospitals and private physiotherapy practices in the community. The aim was to recruit a minimum of three to a maximum of nine children to enable randomization as outlined below. Prior to participation in the study all parents signed an informed consent form and children signed an assent form.

2.3. Procedure

Three factors were incorporated into this SCED design to meet the criteria for Level II scientific evidence. First, the study included *three phases* (Ottenbacher, 2001), which was achieved with an A-B-A design where 'A^1' refers to the baseline (non-treatment) phase, 'B^1' the intervention (treatment) phase and 'A^2' the withdrawal (non-treatment) phase. Second, the onset of treatment was scheduled using a *multiple baseline design* (Kratochwill et al., 2012; Ottenbacher, 2001) which meant that children commenced concurrently and were randomised to a baseline duration of either 5, 6 or 7 weeks. Randomization was performed using concealed random allocation by an investigator not involved in assessment or treatment. Following the baseline period each child participated in 8 weeks of intervention and then 5 weeks of withdrawal. Finally, a *repeated measures design was used with at least 5 data points used throughout each phase* to evaluate stability or variability of the response and trends in the data (Backman et al., 1997; Kratochwill et al., 2012). The minimum number of 5 data points in each phase was chosen to optimize data analysis and minimize the burden on attendance by the family. In addition to repeated measures, a number of measures were collected at selected timepoints and one final assessment of all measures was performed at 3 months following the completion of the intervention. Throughout the study period, children did not receive any physiotherapy or Pilates from other venues but continued their normal sporting activities.

2.4. Pilates intervention

The Pilates intervention was provided in a community based Paediatric Physiotherapy Practice. All children received a dose of 12 h of face to face intervention from a physiotherapist (2 sessions x 45 min × 8 weeks), plus 6 h of a home program under parental supervision (1 session x 45 min × 8 weeks). This dose was designed to meet the basic guidelines for physical activity (Australian Government, 2019) and strength training (Behringer et al., 2010; Lloyd et al., 2014). The 8 weeks in Pilates intervention phase was proposed to be the minimum duration that would allow for the lag time of approximately 6 weeks when strength changes would be expected following introduction of the intervention (Coffey and Hawley, 2007). It was also a pragmatic length for children and families to fit into a school term. The Pilates program was individualised for each child by a physiotherapist experienced in paediatrics (30 years), who was also a trained Pilates instructor (15 years) in the Body Arts and Science International approach (Isacowitz, 2014). The treating physiotherapist was blinded to all assessment results

during the trial.

A detailed description on the Pilates session content and format is provided in the protocol paper for this study (Hornsby, 2021). An example of one Pilates session is provided in Supplementary Material 1. In summary, each session included a structured combination of Pilates exercises performed on a mat and a Reformer (specialised Pilates equipment) at an appropriate difficulty level for each child. The exercises were progressed to a more challenging level of difficulty as the child's strength and endurance improved. At each session, the Pilates instructor recorded exercises performed, the number of repetitions and the amount of resistance used with the reformer (number and weight of springs used to denote light, medium or heavy resistance). Feedback was sought from the child during each exercise to enable modification if any discomfort was felt. Parents were invited to observe each session so they could supervise the individualised written home program of mat exercises performed once per week at home. The parent signed the home program indicating that the home program had been performed and returned it at the next session so adherence could be monitored.

2.5. Outcome measures

Outcome measures are described in detail in the protocol paper (Hornsby, 2021). In summary, outcomes were measured across the International Classification of Functioning, Disability and Health – Children and Youth version (ICF-CY) (World Health, 2007), including (i) pain, (ii) muscle strength, (iii) postural control, (iv) fatigue; (v) physical activity; and (vi) HRQoL. Outcome measures were collected by a physiotherapist who was blinded to the study phases and intervention status. Weekly repeated measures were measured five times during each phase (pain, muscle strength and postural control). Periodic measures were collected twice each phase (fatigue and HRQoL) or at the end of each phase (activity levels). All outcomes were reassessed at 3 months following the completion of the intervention phase to determine medium term retention of changes.

2.5.1. Primary outcome measure

Pain was measured using the PedsQL-Paediatric Pain Questionnaire (PedsQL PPQ) (Sherman et al., 2006; Varni et al., 1987) which is valid and reliable for child self-report and parent proxy-report of pain (Gragg et al., 1996). The Visual Analogue Scale (VAS) scale of the PPQ was completed separately by the child and parent, both reporting on *present pain* and *worst pain in the past week*. Each child then indicated on a body chart their pain locations and intensity (by colour). This data was converted to a scale of 1 (mild), 2 (moderate) or 3 (severe) pain and totalled to calculate the 'cumulative pain intensity' which was then divided by the number of locations to produce the 'mean pain intensity'. Children also recorded written pain descriptors, which were classified under the categories as described by Jensen in 2013 (Jensen et al., 2013) of global pain domains with the subdomains featuring the specific words or phrases used by the children in this study. The number of descriptors each child used in each global pain domain was totalled and the percentage calculated.

2.5.2. Secondary outcome measures

(i) ***Muscle*** strength was quantified using a Hand-Held Dynamometer (HHD) (Lafayette System 01165) which provides the muscle strength readings as peak force in Newtons (N) and is a reliable and valid tool for use in children (Escobar et al., 2017; Hebert et al., 2011). The muscle action groups measured were: hip extensors (gluteus maximus), hip abduction (gluteus medius), knee extension (quadriceps femoris), scapula adduction/downward rotation (rhomboideus major and minor), shoulder external rotation (infraspinatus and teres minor), elbow extension (triceps brachii) and wrist extension (extensor carpi radialis longus, brevis and ulnaris), in the positions described by Daniels and Worthingham's muscle testing manual (Hislop and Montgomery, 2007).

(ii) ***Postural control*** was assessed with 3 items from the Kids-BESTest Balance Evaluation System (Dewar et al., 2017). Two items were selected from the stability limits domain, (i) Functional Reach Test - Forward (FRT-FWD) and (ii) Functional Reach Test - Lateral (FRT-LAT) which were recorded in centimeters. One item was selected from the anticipatory domain, (iii) Stand on one leg (SLS) which was recorded in seconds up to a maximum of 25 s.

Two items were measured approximately monthly as both questionnaires referred to how the child was feeling for the previous month. These were measured in the same week number in each phase for all children, thus accounting for the staggered start of the intervention phase required for the MBD. All questionnaires were completed independently by the children. An initial explanation was given by the blinded assessor to explain the correct procedure and any questions were answered prior to the child completing the measure.

These measures were.

(iii) ***Fatigue*** levels were measured using the PedsQL Multidimensional Fatigue Scale (PedsQL Fatigue) which has demonstrated good validity and reliability for children (Varni et al., 2004). The PedsQL Fatigue includes three domains of: general, sleep/rest and cognitive fatigue. The child is allocated a score of between 0 and 4 points for each question and these scores are then converted to a score of between 0 and 100. The mean overall score is calculated with higher scores an indication of lower fatigue. The minimal clinically important difference (MCID) for the total scores was determined based on data from a similar population (paediatric rheumatology) as 4.9 of the total score for child report and 4.7 for parent report (Varni et al., 2004).

(iv) ***Health Related Quality of Life*** (HRQoL) was measured with the PedsQL Paediatric Quality of Life Inventory (PedsQL 4.0) which has demonstrated good validity and reliability for children with various chronic conditions (Varni et al., 2003). The PedsQL 4.0 includes four domains of physical, emotional, social and school functioning. The child is allocated a score of between 0 and 4 points for each question and these scores are then converted to a score of between 0 and 100. The mean overall score is calculated with higher scores an indication of higher HRQoL. The MCID was used to determine if there is a significant change in the mean scores between each phase. Varni et al., calculated the MCID for HRQoL as a 4.4 change in the total scale score for child self-report and a 4.5 change for parent self-report in HRQoL (Varni et al., 2003).

The final measure was performed in the last week of each phase.

(v) ***Activity*** levels were recorded with an accelerometer (ActiGraph™ wGT3X-BT) which was worn on the hip during waking hours for 7 days in the final week of each phase. The child-parent dyad also documented a daily diary, in which they recorded the times of application and removal of the device and the physical activities that the child performed. To be considered representative, 4 valid days out of 7 are required which includes at least 7 h of data recorded per day and at least one day on the weekend (Voss and Harris, 2017). The average daily times (in minutes) for sedentary, light and moderate vigorous physical activity (MVPA) were calculated.

All outcome measures were repeated at the final 3-month assessment following intervention.

2.6. Data analysis

This study incorporated an A-B-A SCED design, where there was a non-intervention phase preceding and following the intervention phase. Experimental effect was shown in this study if there was a change of greater than 2 standard deviations in the level of an outcome measure when the child commenced the intervention, or when the intervention was withdrawn (Barlow, 2009; Portney, 2020). A hypothetical example of this can be seen in Fig. 1 in the protocol paper for this study (Hornsby, 2021).

Visual analysis of graphic presentation is the commonly used approach to interpret data in a SCED study. In this study, data was plotted against time (Barker et al., 2013) for all the weekly outcome measures including pain, muscle strength and postural control. Evaluation of the data included visual examination for (i) level or magnitude of performance at the start and end of a phase; (ii) trend or direction of change within a phase and (iii) slope or rate of change within the data (Nikles and Mitchell, 2015). Other factors considered were variability of the data in phases, immediacy of intervention effect, data overlap and consistency of data patterns across similar phases (Nikles and Mitchell, 2015). To substantiate visual findings, further statistical analysis was explored using the two standard deviation band method (Perdices and Tate, 2009; Smith, 2012). With this method the mean and standard deviation of data points within each phase were calculated and if at least two consecutive data points in the subsequent phase fell outside the band, then the positive or negative changes were considered significant

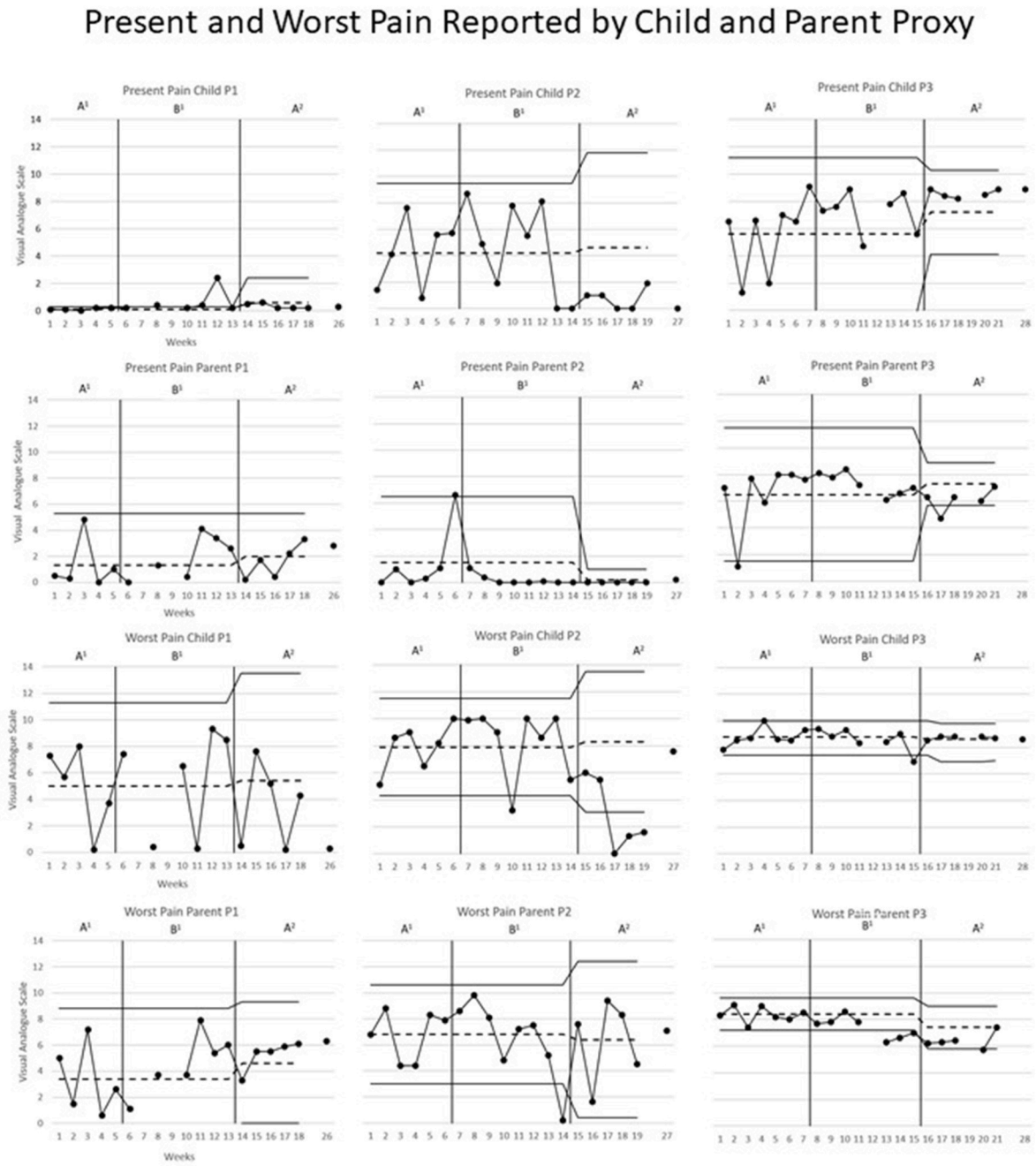

Fig. 1. Visual analysis of present and worst pain for child and parent proxy. A significant increase/decrease is seen if at least 2 consecutive data points are located above/below the 2SD line from the mean for the baseline phase (A^1) to the intervention phase (B^1) or B^1 to the withdrawal phase (A2) in participants (P1: Participant 1; P2: Participant 2; P3: Participant 3). The last data point in A^2 represents the 3 months post intervention result. The bold dotted line represents the mean of the previous phase. The bold solid horizontal lines represents the 2SD lines either above or below the mean.

(Portney, 2020). This method was employed when a participant transitioned from phase A^1 to B^1 and from phase B^1 to A^2. Like visual analysis, these transitions allowed each participant to have two opportunities to demonstrate experimental effect.

For the periodic outcome measures of fatigue and HRQoL, the MCID was used to calculate if significant changes were observed between the different phases. For activity levels, the time was calculated for sedentary, light and MVPA and change in levels were examined between each phase.

3. Results

3.1. Participants

Four children were recruited to the study which included at least one child in each randomization arm of the study. Participant 1 (P1) was a 12 year old male (Beighton Score 6/9) who at the time of recruitment reported chronic pain in both feet, right (R) > left (L), particularly when negotiating stairs and following sport which included basketball and Taekwondo, both bi-weekly. Participant 2 (P2) was an 8 year old female (Beighton Score 6/9) who reported chronic pain in her (R) hip and knee particularly when active and when playing soccer and was swimming bi-weekly. Participant 3 (P3) was a 12 year old male (Beighton Score 6/9) who reported chronic pain in both knees and his hands and who had avoided running, had recently stopped tennis due to pain and still swam bi-weekly. Participant 4 (P4) was an 11 year old female (Beighton Score 7/9) who reported pain in her thoracic spine and both thighs/knees, who was unable to run longer distances for cross country training at school and was involved in ballet 4 times per week. Three of the children commenced in the study concurrently (P1-3). Participant P4 was unable to start until week 2 and then fractured her foot at home during the intervention period, so needed to withdraw from the study.

3.2. Outcome measures

Three outcomes were measured weekly.

3.2.1. Pain

No child showed a reduction in *present pain* or *worst pain* in the past week, during the intervention phase (Fig. 1). Child P2 did show a large reduction in both *present* and *worst pain* towards the end of the intervention which was significant in the early withdrawal phase for *worst pain*, but not for *present pain* as it didn't exceed the 2SD threshold due to wide variation in scores in the baseline phase. At 3 months post intervention, lower *present pain* was maintained for P2. Child P1 showed a drop in *worst pain*. Regardless of the phase, P3 maintained a constant level of *present* and *worst pain*. Parents did not report a reduction in *present* or *worst pain* for their children. There was little similarity

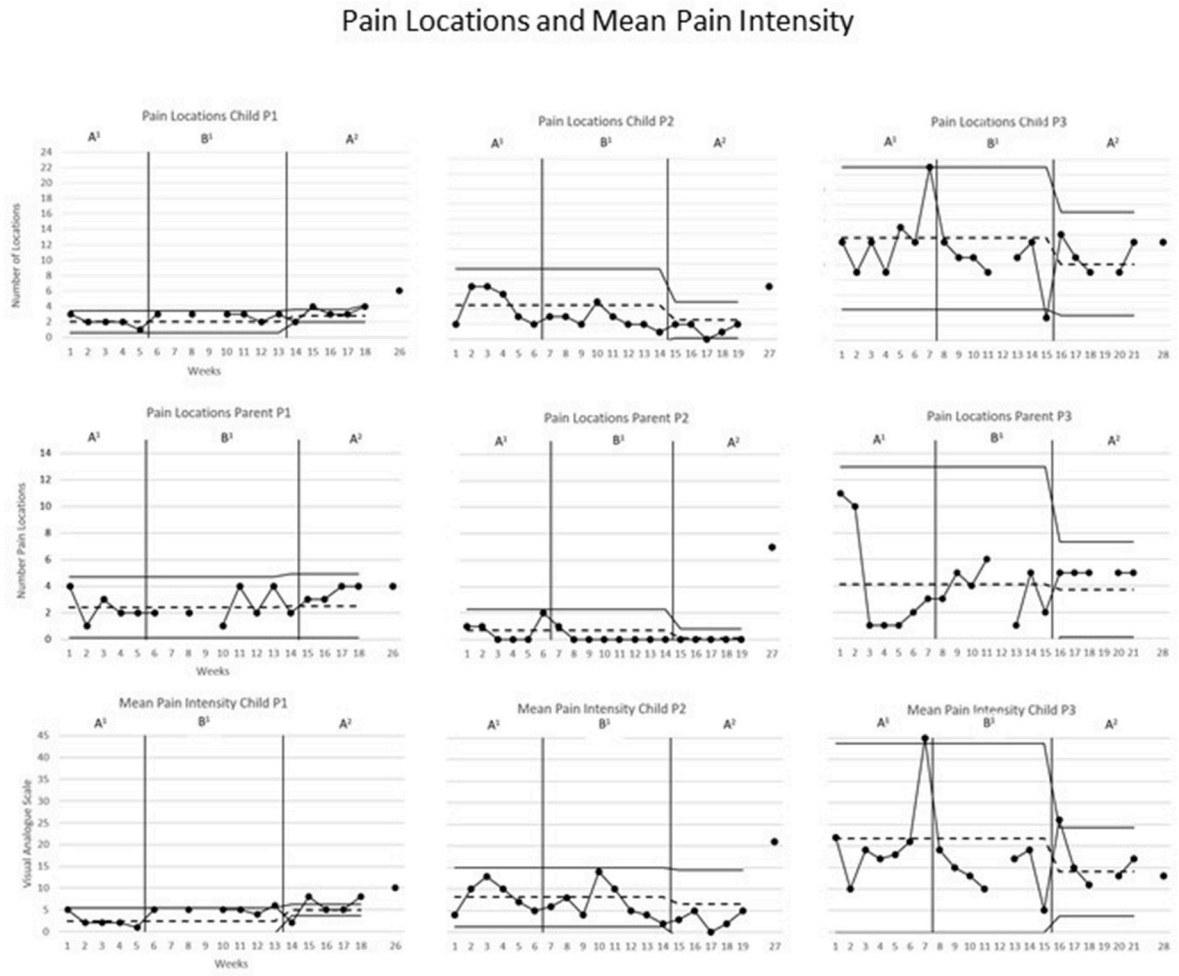

Fig. 2. Visual analysis of pain locations for child and parent proxy. A significant increase/decrease is seen if at least 2 consecutive data points are located above/below the 2SD line from the mean for the baseline phase (A^1) to the intervention phase (B^1) or B^1 to the withdrawal phase (A2) in participants (P1: Participant 1; P2: Participant 2; P3: Participant 3). The last data point in A^2 represents the 3 months post intervention result. The bold dotted line represents the mean of the previous phase. The bold solid horizontal lines represents the 2SD lines either above or below the mean. The mean pain intensity was calculated from the cumulative pain intensity divided by the number of locations.

between the parent-reported and child-reported scores for P1 and P2, with the only similarity seen for P3 for *worst pain*.

The number of pain locations varied according to the child, with the fewest locations reported by P1 (range 1–4), followed by P2 (range 0–7) and the most reported by P3 (range 3–23). No child or their parent reported a significant reduction in pain locations during or following the intervention (Fig. 2). At 3 months post intervention, the number of pain locations increased for P1 and P2 with P1 reaching above the 2SD line of the intervention phase, whilst P3 remained consistent with the mean of the intervention phase. The parent of P2 reported a significant increase in number of locations at 3 months post intervention, whereas the parent of P1 reported no change, and no data was available from the parent of P3. There was some similarity between the number of locations reported by the parent and child of P1, with no similarity seen between the P2 and P3.

Mean pain intensity varied according to the child, with the lowest *mean pain intensity* reported by P3 (range 1.1–1.9), followed by P1 (range 1.0–2.0) and the highest reported by P2 (range 0.0–3.0). No participant reported a reduction in *mean pain intensity* during the intervention or withdrawal phase (Fig. 2) but at 3 months post intervention, intensity had increased significantly for P1 and P2, with no change seen for P3.

The pain descriptors reported by each child are illustrated in Table 1, which is based on the global pain domains described by Jensen in 2013 (Jensen et al., 2013). Each child's choice of words was different with only 1/32 words (ache) being used by all 3 participants and 3/32 used by at least 2 participants (sore, uncomfortable and annoying). Participant 1 used a total of 36 different words in the various domains with 94.4% in the quality domain predominantly using the words 'pressure' and 'pulling' to describe pain, 2.8% of word were from each of the spatial and affect domains. Participant 2 used a total of 43 words with 90.7% in the magnitude domain with 'painful', 'sore', 'hurting' and 'discomfort' being the most used words, with the remaining 9.3% in the quality domain. Participant 3 used nearly double the word count with 83 words which were spread more evenly between the different domains with 51.8% in quality ('burning' and 'searing' being the most frequently used), 18% in temporal, 14.5% in affect, 7.2% in each of magnitude and effect and 1.2% in spatial. Overall, 50% of word descriptors were in the quality domain, 27.8% in the magnitude domain, 9.3% in temporal, 8% in affect, 3.7% in effect and nil in covariate.

Table 1
Pain Descriptors used by Participants (modified Jensen et al., 2013).

Domain Subdomain	Participant 1 Descriptors n = 5	Participant 2 Descriptors n = 15	Participant 3 Descriptors n = 22
Quality (type of pain)			
Ache/aching	1	1	7
Burning/searing			33
Discomfort/Uncomfortable		9	3
Pressure	17		
Pulling/straining	16		1
Stinging		3	
Tense/tight			2
Total	**34 (94.4%)**	**13 (30.2%)**	**46 (55.4%)**
Spatial (where one hurts)			
General pain			1
Long	1		
Total	**1 (2.8%)**	**0**	**1 (1.2%)**
Temporal (when one hurts)			
Everlasting/Long lasting/ Never ending/Ongoing			10
Reoccurring			5
Total	**0**	**0**	**15 (18.1%)**
Magnitude (how much one hurts)			
Low Level: Not very sore/Small pain/Very small amount pain		3	
High Level: Hurt/hurting/pain/ painful/sore		22	2
Intense Level: Extremely painful/very painful/very sore/very, very sore		5	1
Total	**0**	**30 (69.8%)**	**3(73.6%)**
Effect (what pain does)			
Exhausting/Lazy/Tiring			5
Weak			1
Total			**6 (7.2%)**
Affect (how pain makes one feel)			
Annoying/Very annoying	1		10
Distracting			2
Total	**1 (2.8%)**	**0**	**12 (14.5%)**
Covariate (factors associated with pain e.g. activity, position)			
Total	**0**	**0**	**0**
Other	**0**	**0**	**0**
Total			
Grand Total	**36 (100%)**	**43 (100%)**	**83 (100%)**

3.2.2. Muscle strength

Only participant P2 showed a significant increase in strength during the intervention phase with three muscle groups showing two data points above the 2SD line of the baseline phase - left (L) hip abduction and L and right (R) scapula adduction/rotation (Figs. 3 and 4). However, all participants showed a trend upwards in muscle strength in at least two muscle groups just after the end of the intervention phase in the start of the withdrawal phase. These were L hip abduction (P1), L and R knee extension (P1), L and R scapula adduction/rotation (P3), R shoulder external rotation (P2) and R wrist extension (P1 and P2). Scapula adduction and rotation in P2 on both the L and R side were the only muscle actions that demonstrated a significant increase in strength in both the intervention and the withdrawal phase. At 3 months post intervention, muscle groups that had maintained their significant increase in strength were L hip abduction (P2), L and R knee extension (P1), L and R scapula adduction/rotation (P2) and R wrist extension (P1). The graphs for muscle groups that showed significant change for at least one participant are shown in Figs. 3 and 4 with the remainder of the muscle graphs in Supplementary Material 2 (Figs. 5 and 6).

3.2.3. Elements of Postural control

For stability limits in balance, one participant (P1) showed a significant reduction in FRT-FWD distance in the withdrawal phase which was retained at 3 months post intervention, whereas no change was seen for the other two participants. With FRT-LAT, one participant (P3) showed a significant improvement towards the left side in the withdrawal phase which was not retained at 3 months post intervention. No other significant changes were seen in this measure. For anticipatory postural control using SLS, the maximum single leg standing time of 25 s was achieved in all phases on both legs by P3 and late in the baseline phase for P2. This created a ceiling effect where no meaningful analysis could be made. Participant (P1) had some difficulties with balance on one leg with more variability seen for his R leg, but no significant changes were observed. These graphs can be seen in Supplementary Material 2 (Fig. 7).

The range of scores for pain, muscle strength, postural control and balance are displayed in Supplementary Material 2 (Table 3).

Two outcomes were measured twice in each phase.

3.2.4. Fatigue

A reduction in mean total score for fatigue levels, was reported by both P1 and P2 between the baseline to intervention phase, whilst P3 showed an upward trend (Table 2) (. In the withdrawal phase, Participant 3 reported a further increase, whereas P2 reported a further decrease and P1 showed a decreasing trend. All participants maintained their post-intervention fatigue level at the 3 months post intervention

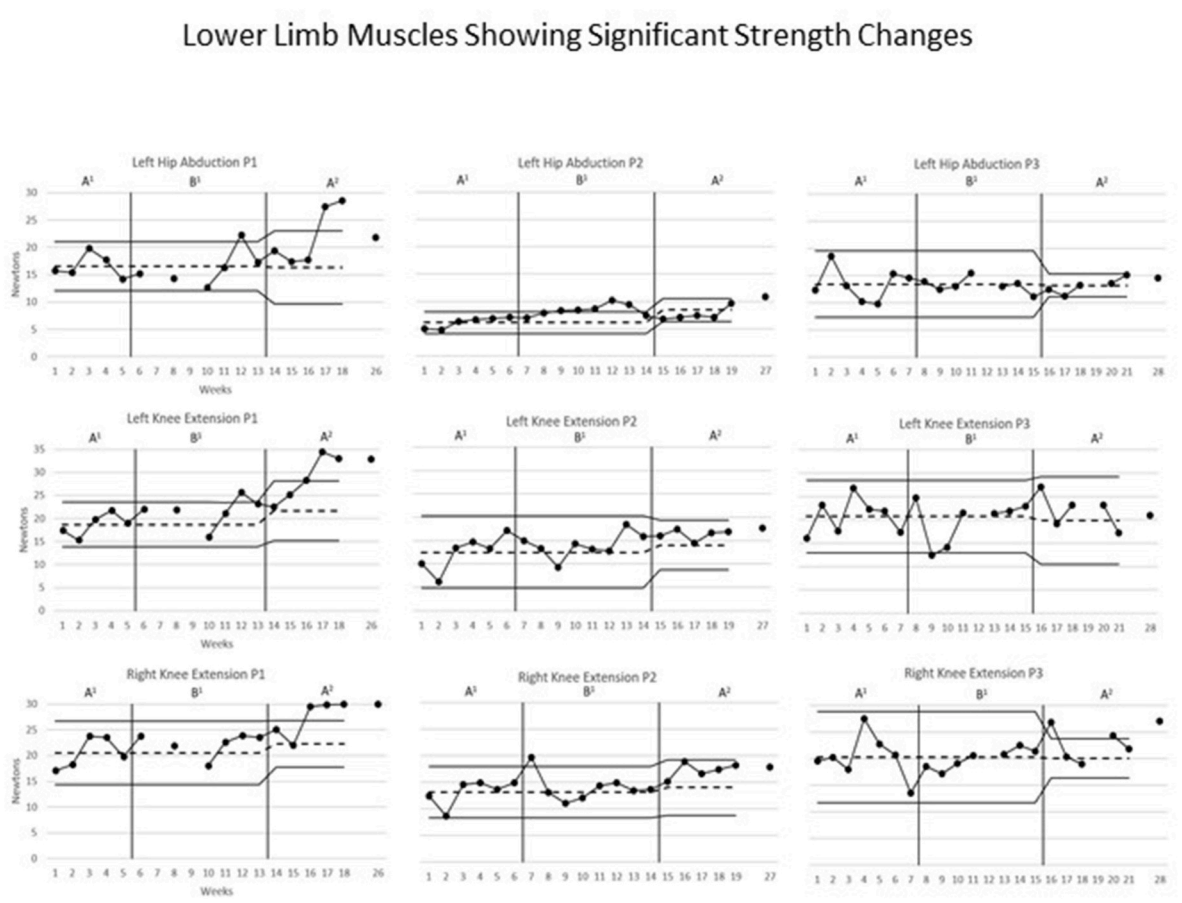

Fig. 3. Visual analysis of muscles in the lower limb which show a significant increase/decrease in strength (at least 2 consecutive data points are located above/below the 2SD line from the mean from the baseline phase (A^1) to the intervention phase (B^1) or B^1 to the withdrawal phase (A2) in a least one participant (P1: Participant 1; P2: Participant 2; P3: Participant 3). The last data point in A^2 represents the 3 months post intervention result. The bold dotted line represents the mean of the previous phase. The bold solid horizontal lines represents the 2SD lines either above or below the mean.

assessment. Parents of P2 and P3 reported reduced fatigue in the intervention phase, which was increased in the withdrawal phase for P3, but maintained by P2. No significant change was reported by the parent of P1 in any phase. At 3 months post intervention, the parents of P1 and P2 reported that the fatigue had been maintained at the same level as the intervention phase and there was no data available for P3.

3.2.5. HRQoL

Participant 1 reported an improvement in the mean total score of HRQoL in the intervention phase, which was maintained through the withdrawal phase and 3 months post intervention assessment. There were no changes in the parent proxy reports (Table 2). Participant 2 and 3 reported no change in HRQoL in any phase in contrast to their parent proxy reports which suggested a marked improvement in the intervention phase for P2 and P3 and again in the withdrawal phase for P3.

3.2.6. Activity levels

Insufficient data available for activity levels meant that no meaningful comparisons between phases could be determined. In the baseline phase, P3 had not worn the Actigraph™ for either day on the weekend so the data was not valid. In the intervention phase, there was a technological problem with downloading the data for P1. In the withdrawal phase, P2 forgot to wear the Actigraph™. The data that was obtained for different types of activity recorded by each child/parent in the daily diary can be seen in Supplementary Material 2 (Table 4).

4. Discussion

This is the first study to use a SCED to examine the impact of a Physiotherapist-led Pilates intervention on pain, strength, postural control, fatigue and HRQoL in children with GJH and chronic musculoskeletal pain. Mixed results were seen across the participants and outcome measures, with clinical benefits for some outcomes and not others. One of the contributing reasons was the inherent variability of some outcome measures during the baseline phase, particularly the primary outcome of pain. This resulted in large standard deviation bands outside of which significant improvement needed to be identified.

4.1. Pain

There was no significant reduction in pain during the intervention phase for any child in this study, however pain did reduce towards the end of the intervention phase for one child and was significantly reduced in the withdrawal phase. This may indicate that a longer period of Pilates intervention is required to achieve pain reduction for this population. This has been seen in two previous Pilates studies that demonstrated decreased pain in children, one with a duration of 3 months (Alves de Araújo et al., 2011) and the other for 6 months (Mendonça et al., 2013). A problem was seen with the self-reported pain pattern with one child, with little change reported in pain even though he had exercised in a pain free range throughout the intervention and progressed with the level of resistance used, number of repetitions and complexity of exercises (Supplementary Material 1). His parent report also indicated a significant decrease in *worst pain* during the intervention

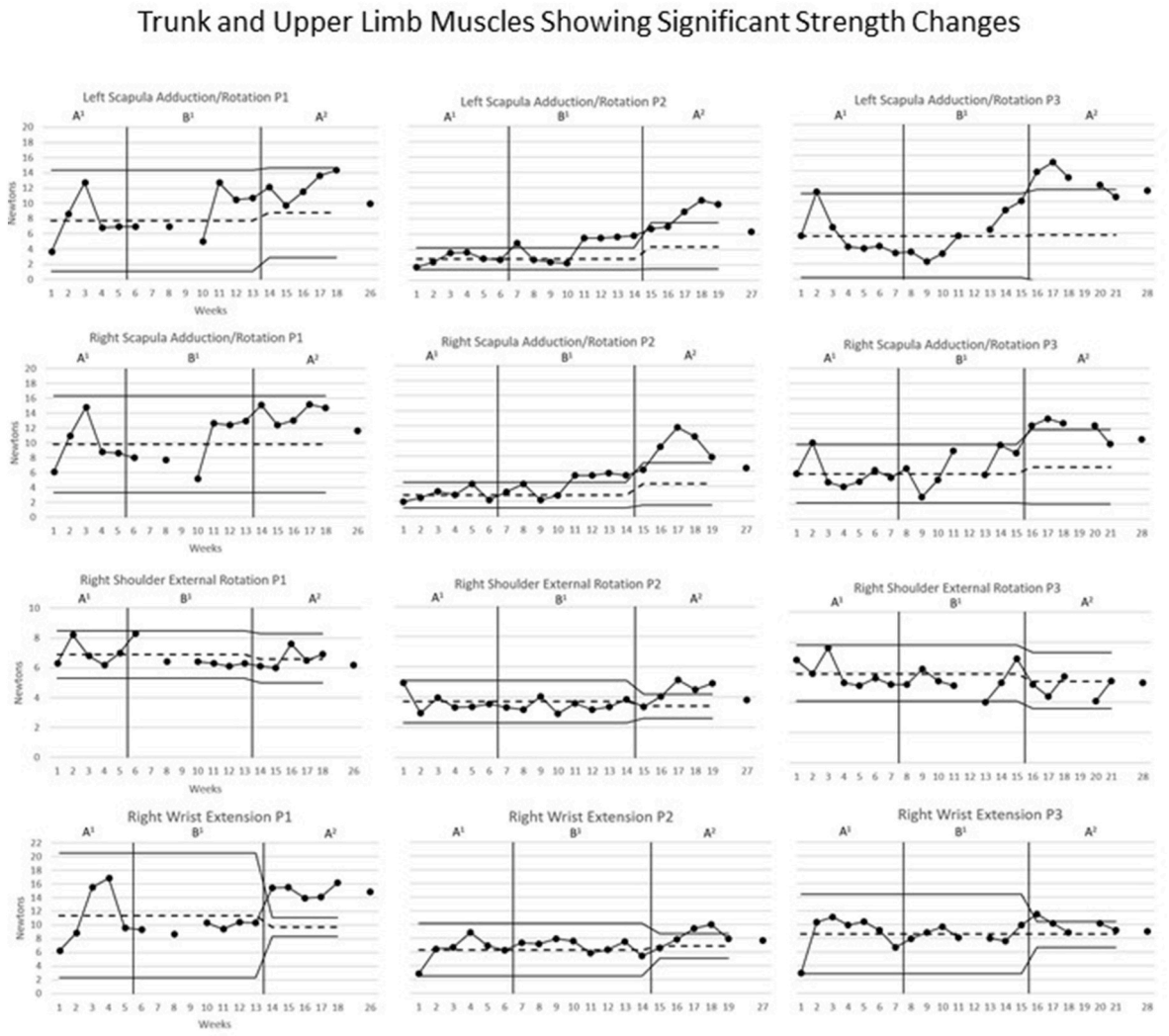

Fig. 4. Visual analysis of muscles in the trunk/upper limb that show a significant increase/decrease in strength (at least 2 consecutive data points are located above/below the 2SD line from the mean from the baseline phase (A^1) to the intervention phase (B^1) or B^1 to the withdrawal phase (A2) in a least one participant (P1: Participant 1; P2: Participant 2; P3: Participant 3). The last data point in A^2 represents the 3 months post intervention result. The bold dotted line represents the mean of the previous phase. The bold solid horizontal lines represents the 2SD lines either above or below the mean.

phase which continued though the withdrawal period. It is possible that this child may have experienced generalised hyperalgesia which impacted pain perception and reporting (Scheper et al., 2015), which requires a multidisciplinary biopsychosocial management approach (Pate et al., 2019).

Importantly, there was no increase in pain reported by any participant during or after the Pilates intervention that was attributed by the children to the intervention. So Pilates has been shown not to produce any added risk as a treatment option (Supplementary Material 2, Table 5). Of note, self-reported pain was highly variable during baseline, intervention and withdrawal phases for most children, due to the impact of daily activities not associated with the intervention. Incidental activities such as participating in the annual school cross country, or falling off a bike on the weekend, caused spikes of pain that were recorded in the pain data but were reported by children to be unrelated to the Pilates intervention.

In future studies, it will be important to consider the way that pain is sampled and reported. We have four main suggestions. First, on the PedsQL PPQ, it is recommended to include additional wording on the questionnaire specifying the cause of pain, to differentiate between intervention versus injury related pain and/or the child's usual chronic pain following activity. Second, each of the areas of pain should be scored separately rather than using an overall body score. This will enable analysis of pain behaviour that is more specific to the intervention provided. Third, to improve utility of Peds QL pain questionnaire, it is recommended that the request for *present pain* is marked more clearly on the body chart page so as not to be confused with *worst pain in the last week*, as on occasions children and parents became confused by this. Fourth, in future studies it would be recommended that more data points are collected in each phase as a pain measure such as the PedsQL PPQ could easily be administered at a similar time daily at home.

Within the SCED design, more data points would hopefully provide more stability in the baseline phase and consequently make visual data analysis results more meaningful. It may also be possible to see variation across the day with pain. This is the first study to include pain descriptors in this population. With further research in larger populations, these descriptors could be standardized for children in various age groups and with different cultural and socioeconomic backgrounds to reflect more accurately the words children use and understand for acute versus chronic pain and pain related to GJH. Finally, it is important in this field to identify valid and reliable parent proxy reports of pain which may be based more on observing a child's pattern and intensity of daily activity than on verbal reports of pain.

Table 2
FATIGUE and HRQoL.

FATIGUE (Higher score indicates less of a problem)						
Participant	Outcome Measure Fatigue	MCID	**Baseline (A^1)** Mean Score Week 1 – (4–6)	Pilates Intervention (B^1) Mean Score Week (8–10) – (12–14)	**Wash Out (A^2)** Mean Score Week (15–17) – (18-/20)	**3 Months Post Intervention** Score Week 26–28
P1 Child	**Total Score**	**4.9**	**59.73**	**69.44 ↑***	**65.28 →**	**69.44 ^→**
P1 Parent	Total Score	4.72	35.42	38.19 →	40.28 →	43.06 ^↑*
P2 Child	**Total Score**	**4.9**	**41.67**	**52.78 ↑***	**47.22 ↓***	**62.50 ^↑***
P2 Parent	Total Score	4.72	65.28	70.14 ↑*	70.14 →	68.06 ^→
P3 Child	**Total Score**	**4.9**	**44.45**	**48.62 →**	**56.25 ↑***	**54.17 ^↑***
P3 Parent	Total Score	4.72	41.67	52.78 ↑*	69.44 ↑*	ND

HRQoL (Higher score indicates better HRQOL)						
Participant	**Outcome Measure** HRQoL	MCID	**Baseline (A^1)** Mean Score	Pilates Intervention (B^1) Mean Score	**Wash Out (A^2)** Mean Score	**3 Months Post Intervention** Score ^
P1 Child	**Total Score**	**4.36**	**70.94**	**76.88 ↑***	**78.29 →**	**80.78 ^→**
P1 Parent	Total Score	4.5	64.3	65.16 →	63.05 →	74.06 ^↑*
P2 Child	**Total Score**	**4.36**	**46.16**	**45.71 →**	**47.43 →**	**62.5 ^↑***
P2 Parent	Total Score	4.5	68.29	69.38 →	76.25 ↑*	83.75 ^↑*
P3 Child	**Total Score**	**4.36**	**51.25**	**53.05 →**	**50.00 →**	**53.13 ^→**
P3 Parent	Total Score	4.5	56.33	64.15 ↑*	71.02 ↑*	ND

Abbreviations: A^1: Baseline Phase; A^2: Pilates Intervention Phase; B^1: Withdrawal Phase; ND: No Data; MCID: Minimally Clinically Important Difference - calculated by SEM and derived by multiplying the SD by the square root of 1-alpha (Cronbach alpha reliability coefficient, Varni et al., 2003 for HRQoL/Varni et al., 2004 for Fatigue); ↑*: Significant improvement from the previous phase; ↓*: Significant decrease from previous phase; → Maintained level from previous phase; ^: The 3 months post intervention score was compared to the mean score from the Pilates Intervention Phase.

4.2. Muscle strength

As for pain there was a trend for muscle strength to increase towards the end of the intervention phase in at least some muscle groups for all children. This improvement continued or was mostly maintained for P1 and P2 for much of the withdrawal period and on occasions for P3 (Fig. 3). Slow improvements in muscle strength and maintenance of strength for a period after intervention can be a feature of exercise interventions for children, especially children with hypermobility (Simmonds and Keer, 2007), which is different to interventions that involve medications where a more immediate result is expected. A lag time of approximately 6 weeks has been reported for improvements in strength after introduction of intervention (Faigenbaum et al., 1996, 1999), with maintenance up to 4 weeks following withdrawal (Faigenbaum et al., 1996; Fleck, 1994). With this in mind, the philosophy of Pilates and the needs of the hypermobile population are aligned, which are to exercise in a pain free range rather than rapidly introduce increasing resistance. Pilates encourages a more gentle and gradual approach to developing strength and motor control (Latey, 2002). Therefore, it is recommended that future studies involving children with GJH and chronic musculoskeletal pain, involve a longer intervention period of at least 16 weeks to give sufficient time to demonstrate change. Even more ideal would be to conceptualise and provide Pilates access as a regular, safe physical activity.

Review of treatment records during the intervention phase of this study indicated that children improved in their level of resistance of the springs used for exercises and in the difficulty level of the Pilates exercises performed. An example of a Pilates session and the number of springs used for a particular exercise can be seen in Supplementary Material 1. This shows that the children were improving in their strength and control of movement and these observations are possibly a better indicator of strength than manual muscle testing via HHD for this population. At times, a child had difficulty performing a manual muscle test for a particular muscle group due to pain and this may have contributed to the variability of strength results seen. Previous studies reporting improvement in strength following Pilates included dancers (Amorim et al., 2011) and children with Diabetes Mellitus (Tunar et al., 2012), but neither population included children impacted by pain. There were two studies involving children with chronic pain. One study involved children with Juvenile Idiopathic Arthritis (Mendonça et al., 2013) and the other non-structural scoliosis (Alves de Araújo et al., 2011) which assessed pain but these studies did not examine muscle strength. Future studies could include measures of functional exercise performance as well as formal muscle testing to track the impact of pain. Alternatively, functional measures of muscle strength such as the vertical jump and seated throw may provide more useful information and may not be as confounded by pain.

The original intent of this study was to demonstrate an increase in strength and control over a period spanning eight weeks. As it now appears to require a longer duration, there was not much opportunity to demonstrate this. As the basis of practicing Pilates exercise is to recruit core muscles such as the transverse abdominus and pelvic and scapula stabilisers, this aim was supported in the finding that improved strength was particularly seen in scapular adduction/downward rotation and hip abduction muscle groups. Another possible explanation for scapula control showing the most significant improvement, could be that it is a muscle group that is commonly weaker in more sedentary individuals, so there may have been more opportunity for improvement than other muscle groups (Faigenbaum et al., 1996).

4.3. Dose of intervention

Even though good compliance was documented with the twice weekly Pilates intervention at 75–100% within the clinic and 62.5–75.5% for the once weekly home program, this study may have been underdosed at eight weeks of intervention and require a longer duration. Circumstances did arise where some participants were unable to attend a session either due to illness, school events, or unplanned family holidays which coincided with the intervention period, so some of the intervention time was lost. This needs to be allowed for in the minimum dose in future studies. A suggested increase to at least 16 weeks of intervention would compensate for the slower response seen for muscle strength, motor control and other physical changes. This will allow for any missed sessions but still involve a duration that is realistic for families to manage (Faigenbaum et al., 1996).

4.4. Elements of postural control

Three of the balance evaluation items in the Kids-BESTest (Dewar et al., 2017) were examined. Stability limits was measured with FRT-FWD which did not show improvement, but the FRT- LAT did show improvement for one participant in the early withdrawal phase. One possible explanation could be that all participants had pain in the lower limbs which could inhibit the ability to reach or that many Pilates exercises did not provide sufficient postural control demand for limits of stability in standing e.g., mat or reformer work in the supine position. In anticipatory postural adjustment, the item of SLS up to 25 s reached a ceiling effect in the baseline phase for 2/3 participants, so was not possible to show change for this more capable population. A more challenging test of limits of stability could have been the Star Excursion Balance Test. This would be suitable to examine outcomes of Pilates which have been shown to improve dynamic balance (McMillan et al., 1998) more so than static balance (Park et al., 2016).

4.5. Fatigue

As fatigue is one of the factors commonly reported with GJH (Simmonds et al., 2019), it is notable that there was a significant improvement in fatigue levels for 2 participants in the intervention phase with the third participant showing an upward trend which became significant in the withdrawal period. This is supported by a cross-sectional study by Scheper in 2017 (M. C. Scheper et al., 2017; M. Scheper et al., 2017), who also measured fatigue in this population and found higher pain intensity was associated with lower muscle endurance, postural control and higher fatigue along with more severe functional impairment. In our study, there was poor alignment in fatigue scores between children and their parents, so we would recommend that a proxy report not be used instead of a child report.

4.6. Quality of life

Pain and fatigue have been associated with reduced HRQoL in children with GJH and chronic musculoskeletal pain (Fatoye et al., 2012; Pacey et al., 2015). In this study only one child and one parent of a different child reported improved HRQoL in the intervention phase and two parents in the withdrawal phase. One child and parent agreed on the HRQoL level in both the intervention phase and 3 months post intervention. A different child and parent agreed in the withdrawal phase. In contrast, the parent and child reported HRQoL scores were strongly correlated in the Pacey study in 2015 (Pacey et al., 2015). However this involved a much larger sample (n = 89) and was not associated with intervention. Parent insight into the HRQoL experience of children is important as they are the ones who present their child for medical investigation. However a proxy report cannot be assumed to replace a child report. Further research in both self-reported and proxy reporting of HRQoL is recommended. In future research it would beneficial to not only analyse the total scores of the Peds QL 4.0 but the subdomains to give more insight into this population and effect of the intervention.

4.7. Outcome measures

In addition to the outcome measures mentioned above, measures that specifically assess engagement in physical activity and qualitative perceptions of participation may provide further insight into a child's view of the Pilates intervention. In particular future research should include the identification of a child selected goal using the Goal Attainment Scale (GAS) to be assessed before and after intervention.

4.8. Study limitations

There were a few limitations to the outcomes of this study. First, although some significant results were reached, it was difficult to prove significance for some outcome measures (especially pain) due to the high variability in these measures in the baseline phase. For future SCED studies for this population, it is important to find an outcome measure that is more stable, such as engagement in daily activity or extend the timeline of the monitoring phases to attempt to reach stability. Second, although parents and children completed questionnaires as specified in instruction manuals, there was some missing data, sometimes children or parents were confused by multiple pain questions on the PedsQL PPQ, or some children forgot to wear their ActiGraph™. Consideration of child friendly outcome measures is important for this population such as strength measured directly from Pilates equipment. Finally, pain and fatigue caused from general activity was highlighted under 'other incidents' in the adverse events table (Supplementary Material 2, Table 5) with multiple incidents seen that triggered a temporary spike in pain or fatigue that were not related to the Pilates intervention. As these additional incidents may have impacted the results, it would be recommended to limit unusual activity that the child was not adequately prepared for (such as school cross country events), to gain a clearer indication of the potential benefits from an intervention.

5. Conclusion

This study was the first to evaluate the impact of a Physiotherapist-led Pilates intervention using a SCED design in school aged children with GJH and chronic musculoskeletal pain. Outcomes were measured across the ICF including pain, strength, postural control, fatigue and HRQoL. Results were variable for this eight-week intervention period but some positive improvements were seen in reducing fatigue and possible benefits for improving muscle strength and HRQoL. Limited conclusions can be drawn regarding pain and postural control. Further research is recommended to confirm if a longer duration of intervention is more effective. Future research could incorporate outcome measures that more specifically measure functional strength, engagement with physical activity and achievement of participation goals. A longer follow up period may confirm sustainability of benefit. Nevertheless, this study adds to a growing body of evidence of the effect of Pilates for children with musculoskeletal and/or pain conditions and may provide therapists with an additional management option. This study may underpin future intervention studies involving larger numbers of children in a SCED or randomised control trial.

CRediT authorship contribution statement

Elizabeth A. Hornsby: Conceptualization, Formal analysis, Investigation, Methodology, Project administration, Visualization, Writing – original draft. **Leanne M. Johnston:** Conceptualization, Methodology, Supervision, Writing – review & editing.

Declaration of competing interest

The authors state that they had no interests that could be perceived as posing as a conflict or bias.

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Appendix A. Supplementary data

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