Effect of Sitting Posture on Development of Scoliosis in Duchenne Muscular Dystrophy Cases

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ABSTRACT

Background: Scoliosis is a frequent association in boys with Duchenne Muscular Dystrophy when the ability to walk is lost around nine to 12 years of age. This study assessed the contribution of physical factors including lumbar posture to scoliosis in non-ambulatory youth with DMD in Nepal.

Methods: Linear regression was used to assess effects of time since loss of ambulation, muscle strength, functional severity and lumbar angle as a binary variable on coronal Cobb angle; again logistic regression was used to assess effects of muscle strength and cross-legged sitting on the presence of a lordotic lumbar posture in 22 non-ambulant boys and young men.

Results: The boys and young men had a mean (SD) age of 15.1 (4.0) years, had been non-ambulant for 48.6 (33.8) months and used a median of 3.5 (range 2 to 7) postures a day. The mean Cobb angle was 15.1 (range 0 to 70) degrees. Optimal accuracy in predicting scoliosis was obtained with a lumbar angle of ≤-6° as measured by skin markers, and both a lumbar angle ≤-6° (P=0.112) and better functional ability (P=0.102) were associated with less scoliosis. Use of cross-legged sitting postures during the day was associated with a lumbar angle ≤-6° (OR 0.061; 95% CI 0.005 – 0.672; P=0.022).

Conclusions: Use of cross-legged sitting posture was associated with increase in lumbar lordosis. Higher angle of lumbar lordosis and better functional ability are associated with lesser degree of scoliosis.

Keywords: Duchenne Muscular Dystrophy; lumbar lordosis; lumbar posture; Nepal; scoliosis.

INTRODUCTION

Duchenne Muscular Dystrophy (DMD) is a X-linked recessive disorder due to mutation in the dystrophin gene,1 that cause progressive muscle weakness leading to feet off around 9 to 12 years of age and scoliosis often develops.2 Different treatment options are recommended to prevent scoliosis1 however, it does develop in many boys.4 Surgery is considered for moderate to large curves, ideally before respiratory function deterioration.5, 6 The needs of DMD boys in Nepal are now being acknowledged and rehabilitation strategies used proactively.7 It has been observed that they have prolonged ambulatory phase,8 and scoliosis is not common even after loss of ambulation.8 Culturally, Nepalese people practice cross legged floor sitting. In DMD boys with muscle weakness, this posture may increase lumbar lordosis that has been associated with greater spinal stability.10 In this study, we sought to examine and identify the relationships between cross legged sitting, lumbar posture and scoliosis.
METHODS

Twenty five boys and young men with DMD from Nepal at non-ambulant stage who had not received surgical interventions were included in this study, after taking verbal and written consent from the boys and their parents. Relevant information such as age when ability to walk was lost was reported by the primary caregiver or the client himself.

Each case was examined for muscle strength, functional ability, lumbar sitting posture, spinal deformities, posture through the course of the day, as detailed below. Finally, past and current use of glucocorticoid medications was recorded as a binary variable.

1. Muscle strength was assessed with manual muscle testing, each muscle group was graded using the Medical Research Council (0 to 5) scale which has been found reliable in DMD. A total muscle strength score was calculated by dividing the sum of the individual muscle group scores by the total possible muscle strength score and expressing as a percentage.

2. Functional abilities were measured with the EK scale, a functional severity scale developed specifically for non-ambulatory boys and men with DMD giving a score ranging from 0 to 30, and scores have explained variation in muscle strength, contractures and respiratory capacity.

3. Lumbar posture was assessed with a lateral photograph of the lumbar spine taken whilst seated on a firm-surfaced stool with hips and knees at 90 degrees flexion, and feet as flat as possible. Photoreflective markers were taped (3M doubled sided tape, Pymble, NSW) to the bony landmarks of the right anterior superior iliac spine, midpoint of the greater trochanter, lateral femoral condyle, lateral malleolus and the spinous processes of T10, L2, L4 and S2. A camera (Canon EOS 500D) was placed on a tripod a distance of 1.5m away and at the level of T8. Sitting posture as tall as possible (with minimal support as necessary for safety) was then photographed. A lumbar angle was calculated as the angle between the intersection of the tangents drawn through the T10/L2 markers and the L4/S2 markers and measured with Image J software, a Java image processing program that measures angles between manually marked positions on a digital image (National Institute of Mental Health, Bethesda, Maryland, USA) (Figure 1a). This measure was applicable to in-the-field measurement. A lateral spinal X-ray was taken in side lying position and lordosis was measured using standard methods (Figure 1b).

4. Scoliosis was assessed clinically and an antero-posterior X-ray of the spine was taken in supine. Any lateral curvature in spine was measured as the Cobb angle using standard methods (Figure 1c).

5. Postures over the course of one day were recorded by the primary caregiver on a 24 hour diary card with response categories indicating the type of lying, sitting and standing used during each 15 minute epoch of the day.

This study was approved by the Ethics Committees of the Nepal Health Research Council, Kathmandu, Princess Margaret Hospital and Curtin University, Perth.

RESULTS

All 25 boys and their families who were eligible for the study agreed to participate although three could not have an X-ray due to geographical barriers, and the findings of 22 DMD boys are therefore reported. The average age was 15.1 ± 4.0 years and the boys had been non-ambulant for 48.6 ± 33.8 months. The average MRC muscle strength score was 39.7% ± 12.1 and the average EK functional severity score was 12.5 ± 5.6. Older age
(coeff 0.68, P=0.022), increased time since walking was lost (coeff 0.11, P=0.001) and weaker muscle strength (coeff -0.33, P=0.001) predicted poorer EK severity scores. As is usual for DMD, the older boys in our sample were more severely affected reflecting increasing weakness with time. Five (22.7%) of the group had previously used and 13/22 (59.1%) were currently using glucocorticoid medications.

The DMD boys were awake an average of 16.9 (1.0) hours and adopted a median of 3.5 (range 2 to 7) different postures during the day. Supine lying (n=16; 72.7%), side lying (n=18; 81.8%) and cross legged sitting (n=15; 68.2%) were common postures during the course of their usual day. Other postures included diamond sitting on the floor and sitting on a chair (Table 1). No boy sat in a wheelchair with postural supports or was supported for standing.

The postural lumbar angle during stool sitting was on average -3.6 (13.3) degrees, ranging from 29.8 to -17.8, three of the young men needing minimal support to maintain balance for their photograph. The area under the ROC curve was 0.767 (95%CI 0.555-0.978) for lumbar angle as a predictor of the presence of scoliosis (>10°), and optimal accuracy (taking into account sensitivity and specificity) was obtained with an angle of -6° (Figure 2). The lumbar angle increased with the size of the lordosis as measured by lateral X-ray taken in side lying (coefficient 0.738; 95% CI 0.227-1.249; P=0.007). A photographically derived angle of ≤-6° was observed in 12/22 (54.5%). The mean Cobb angle was 15.1° (range 0 to 70°): four boys aged 11, 16, 17 and 19 years had a coronal Cobb angle ≥30°. Three of 12 who were 15 years or older (25%) and two of eight who were 17 years or older (25%) had a curve ≥30°.

Table 1. Different postures used over the day by 22 non-ambulant boys and young men in Nepal.

<table>
<thead>
<tr>
<th>Posture</th>
<th>N (%)</th>
<th>Mean (SD)</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supine lying</td>
<td>16 (72.7)</td>
<td>2.6 (1.2)</td>
<td>0.8-5.0</td>
</tr>
<tr>
<td>Prone lying</td>
<td>6 (27.3)</td>
<td>0.9 (0.5)</td>
<td>0.3-1.5</td>
</tr>
<tr>
<td>Side lying</td>
<td>18 (81.8)</td>
<td>3.2 (1.6)</td>
<td>1.5-7.0</td>
</tr>
<tr>
<td>Stool</td>
<td>3 (13.6)</td>
<td>2.8 (3.1)</td>
<td>0.8-6.3</td>
</tr>
<tr>
<td>Floor sitting</td>
<td>6 (27.3)</td>
<td>4.4 (1.9)</td>
<td>1.8-6.5</td>
</tr>
<tr>
<td>Chair</td>
<td>10 (45.5)</td>
<td>6.3 (3.1)</td>
<td>1.5-11.3</td>
</tr>
</tbody>
</table>

*average and range statistics calculated for those who used the posture; *wheelchair without additional seating supports

| Time off feet | 0.14 (0.11 - 0.38) | 0.258 | 0.051 (0.00 - 0.33) | 0.159 |
| MRC%          | -0.51 (-1.18 - 0.17) | 0.131 | -0.43 (-1.12 - 0.26) | 0.211 |
| EK severity score | 1.19 (-0.26 - 2.64) | 0.102 | 0.86 (-0.65 - 2.37) | 0.102 |
| Lumbar angle (<6° vs ≥6°) | -13.9 (-29.52 - 1.72) | 0.078 | -10.75 (-27.22 - 5.72) | 0.012 |

All cases (n=22) Excluding case with lumbar lordosis and kyphosis (n=21)

<table>
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<tr>
<th>Coefficient (95% CI)</th>
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<td>-10.75 (-27.22 - 5.72)</td>
</tr>
</tbody>
</table>

**Table 2. Relationships between predictor variables and coronal Cobb angle calculated with linear regression, for all cases (n=22) and without one case who presented with both lumbar kyphosis and lordosis (n=21).**

*average and range statistics calculated for those who used the posture; *wheelchair without additional seating supports

Age and time since the ability to walk was lost did not predict coronal Cobb angle. The EK severity score (reflecting functional ability) and a lumbar angle ≤-6° were stronger predictors with the latter being the strongest univariate predictor (coefficient -13.9, P=.078) (Table II). Figure 3 shows a lateral photograph of a 15 year old man with a lumbar angle ≤-6° and whose Cobb angle was 10°.
DISCUSSION

Duchenne Muscular Dystrophy is characterized by progressive muscle weakness which is due to the disease process itself and also secondary to disuse atrophy. Joint contracture and development of deformities including that of spine is due to lack of support from weak muscles. Spinal deformity, scoliosis, usually develops after loss of ambulation. In DMD boys living in Nepal, we found that a typical day involved time spent in supine and side lying posture, sitting in an unadapted wheelchair and/or sitting in cross legged sitting on the floor. We identified a weak relationship between a lordotic lumbar posture in sitting and a less severe scoliosis, with this lumbar posture more likely when cross-legged sitting postures were used during the day. The boys and young men in our sample lost muscle strength and function as would be expected in DMD, but there was only a weak relationship between the size of the Cobb angle and time since they had lost the ability to walk. As would be expected, the relationship between the lumbar sitting posture and Cobb angles was confounded by functional severity but a lumbar angle ≤-6° was nevertheless associated with approximately 11° reduction in Cobb angle. Therefore, use of a variety of posture daily including those that encourage a lordotic lumbar posture could be protective against scoliosis.

One fifth of our sample had a Cobb angle of more than 30°, this is significantly (P<0.004) lower than in a longitudinal clinical review of 123 boys to the age of 17 years in the UK showing a little more than three quarters developed a similarly severe scoliosis. Use of daily glucocorticoid medications over the long term is
associated with delayed onset of scoliosis as well as its slower development, likely related to slower decline in muscle strength. Approximately 30% of the UK cohort had received prednisolone according to local protocols but a moderate to severe scoliosis nevertheless developed for many. In our sample, past or current glucocorticoid use was not associated with Cobb angle although their duration of use had in many cases been relatively short.

For those in Nepal, economic factors reduce opportunities for the implementation of comprehensive management, and we would expect poorer outcomes but the proportion who had developed scoliosis was much smaller than expected. It is interesting also to note a prolonged ambulatory phase among Nepalese DMD boys compared to that of the Japanese boys.

A variety of postures were used daily including cross legged sitting on the floor. This is a widely practiced sitting position in Nepal over long periods of time throughout the day and its practice was associated with maintenance of a lordotic lumbar angle. The ability to sit cross legged on the floor with a lumbar lordosis requires adequate range of movement at the lumbar spine and hip joints in order to maintain anterior rotation of the pelvis in combination with flexion, abduction and external rotation at the hip. With poor muscle strength as seen in DMD, the upper trunk is then held high and balanced over this base of support. This possibly reduces axial rotation of the spine and presumably its contribution to a developing scoliosis. A long term environment that includes practice of cross legged sitting may allow development of the necessary range of motion at the pelvis and hips to achieve this position, even before the boys lose the ability to walk.

With muscle weakness, the spine often develops a long thoracolumbar scoliosis in the first instance that later becomes kyphoscoliotic; and the presence of a lumbar lordosis has been suggested as protective of spinal symmetry in DMD. For example, Oda et al found seven of 43 patients with DMD who did not have progressive spinal deformity to have normal sagittal plane alignment and a scoliosis of <30°. In a laboratory study, a lumbar lordosis when sitting on a base tilted 15° was associated with better tolerance of spinal loads in eight non-ambulant boys with DMD. For our sample, maintaining a lumbar angle -6° as measured photographically was the strongest univariate predictor of Cobb angle, more so than factors such as time since walking was lost and muscle strength, and also despite the absence of supported seating systems. The effect of lumbar angle was moderated by functional severity in the multivariate model, as would be expected, and this effect was stronger when the case with an unusual deformity of both lumbar kyphosis and lordosis was excluded. Using multiple postures throughout the day including cross legged sitting may be protective of spinal symmetry, although we recognize that these strategies are not directly applicable to boys in different cultures.

To characterize lumbar lordosis, we used a field measure to examine lumbar posture with skin markers and found that a lumbar angle -6° was the cut-point with optimal sensitivity and specificity for predicting the presence of scoliosis. The boys had lean body shapes and we were confident that the skin markers were able to illustrate the posture of the lumbar spine. Data were available to assess the relationship between radiological measurement of lumbar lordosis in side lying and our photographic measure for some of our sample, and the strong relationship was encouraging. We elected to use the photographic measure in our analyses because it was assessed in the sitting position and its in-the-field application. Further study of this measure taking account of any influence of BMI would be valuable to understanding its application to other populations.

Our study is the first to report physical assessment of DMD boys in a developing country and our findings are particularly applicable to those living in similar countries. We also feel that assessment of boys living within different environmental circumstances to those participating in most other studies in the literature affords a valuable opportunity to learn much more about factors that can influence outcomes. Our recruitment rate was high and we assessed a comprehensive range of variables to describe impairments and activities. However, we also acknowledge the limitations of our study. The sample size was unavoidably small comprising boys and young men of families who lived in the Kathmandu area and we were interested to receive interventions for their son by being registered with the Muscular Dystrophy Foundation Nepal. Only cross-sectional data was collected and we would recommend additional longitudinal study to track this potential protective role of sitting habit.

Our data suggest that non-ambulant boys and young men with DMD in Nepal are less likely to develop scoliosis in comparison to those reported in the literature, they utilize multiple postures throughout daily life and lumbar postures characterized by lordosis afforded modest protection of coronal symmetry of the spine. Continued monitoring of scoliosis in DMD in the literature to understand spinal postures in relation to evolving glucocorticoid regimens and physical management strategies is important in a variety of cultural settings, and we argue for greater investigation of neuromuscular disability in majority world countries. Support for spinal and pelvic symmetry as well as spinal extension is recommended to be built into wheelchairs for those who
are non-ambulant, but specific guidelines for the lumbar spine have not been found. Development and testing of seating strategies that allow for greater postural variety including reproduction of a sitting posture with the lumbar angle at approximately -6° could confer additional advantages for non-ambulant boys with DMD.

CONCLUSIONS

Use of cross-legged sitting posture was associated with increase in lumbar lordosis. Higher angle of lumbar lordosis and better functional ability are associated with lesser degree of scoliosis.

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We would like to acknowledge the valuable contributions of the boys and young men with Duchenne Muscular Dystrophy in Nepal and their families. We are also grateful for the support of the staff and Board members of the Muscular Dystrophy Foundation Nepal, the Muscular Dystrophy Child Care Society Nepal and the Muscular Dystrophy Association Nepal.

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