Gower’s sign in children with juvenile idiopathic arthritis

Sir, One clinical sign assessing proximal leg muscle weakness is Gower’s sign, the most important feature of which is the adoption of a prone position before standing. This can be seen during normal development in toddlers up to the age of 36 months; only 6.5% of healthy children over 3 yr still roll prone during standing, while children with neuromuscular disorders have a positive early Gower’s sign that persists after the age of 3 yr [1]. Previous research in our department [2] found that 58% of children with juvenile idiopathic arthritis (JIA) older than 7 yr had difficulty standing from the floor. This was the commonest area of difficulty in the JAFAR-C (Juvenile Arthritis Functional Assessment Report for Children) and JAFAR-P (Juvenile Arthritis Functional Assessment Report for Parents) [3], a functional tool used in the assessment of children with arthritis. The aim of this observational study was to evaluate the presence of an early Gower’s sign in children with JIA, and find the correlated factors.

Children with JIA attending the regional paediatric rheumatology clinic for follow-up were asked to lie supine on the floor and then to stand as fast as they could three times. Results were analysed according to whether the child adopted a prone position on at least two occasions at any time during standing, or showed difficulty rising from the floor. Following parental consent we obtained from the notes the complete history of onset, subsequent progression, symptoms, treatment and complete charting of the previous affected joints, as well as the global disease activity at the time of the study of active disease or of disease in remission. The parent was asked to complete a JAFAR-P questionnaire modified by an additional question to a total of 24 items [2]: ‘Over the past week how often has your child been able to perform the following activity ‘Get up from floor without holding on’: all the time/sometimes/almost never’.

Thirty children with JIA were assessed: 19 girls and 11 boys, aged from 3 yr to 16 yr 8 months (mean 9 yr 4 months). At the time of the study the disease was systemic in one patient, polyarticular in 13 patients and pauciarticular in 16 patients. The age of onset of the disease ranged from 18 months to 15½ yr (mean 5 yr 4 months). Ten children required treatment with methotrexate.

The pattern and ease of standing from the floor was normal in 14 children (47%), three children (10%) failed to stand from the supine position on all three attempts, and six (20%) showed a positive early Gower’s sign by rolling prone, kneeling and pushing with both hands.
when attempting to stand on at least two occasions. Seven (23%) showed an abnormal pattern of standing and moderate difficulty, by rolling prone, kneeling and pushing with one hand to stand or rolling prone, without kneeling, and pushing with both hands. In total, 53% of the children with JIA in our study had difficulty getting off the floor, 10% failing to stand.

Table 1 shows that children with active JIA, the poly-articular type of the disease, and active involvement of the hip during assessment or in the past and active involvement of the knee during assessment had more difficulty in standing from the floor. All patients with active hip involvement had a positive Gower’s sign. One of the children who failed to stand had contractures of both hips and another one with a positive early Gower’s sign had contractures of both knees. The JAFAR-P score showed a correlation with difficulty standing off the floor; it was low in the group of children with negative Gower’s sign, increased as the abnormal pattern of standing got worse, and was highest in the group who failed to stand.

Our results suggest that JIA may cause proximal leg muscle weakness, resulting in a positive early Gower’s sign. Research in adults with rheumatoid arthritis showed decreased isometric and isokinetic muscular strength [4], mainly in the early phase of the disease, but progressive dynamic strength training rapidly increased the neuromuscular performance capacity to the level of healthy people [5]; the gain in performance was lost to a great degree during the detraining period [6]. On the other hand, Gower’s is a non-specific sign and, as with other tests of muscle strength in children with arthritis, it may be possible that the arthritis itself causes the difficulty in rising from the floor.

In conclusion, the early Gower’s sign is a useful part of the clinical examination of children with JIA, identifying possible muscle weakness of the relevant joints. Children cooperate well in performing the task of standing up from the floor, as they do this anyway many times during the day, while all other methods of assessing muscle power or joint mobility require a greater degree of cooperation from the child.

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