Validation of Beighton Score and Prevalence of Connective Tissue Signs in 773 Dutch Children

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ABSTRACT. ***Objective.*** Validation of the Beighton Score and the prevalence of connective tissue signs were investigated in Dutch children.

Methods. Hypermobility investigation according to Beighton was undertaken in 773 healthy children aged 4–12 years. An inventory of the signs that fitted with connective tissue disorders was compiled.

Results. The percentage of general hypermobility at a cutoff point of ≥ 4 was 26.5% (range 11.4–49%) in children aged 4–9 yrs. At the age of 10–12 yrs, this percentage was 5.3% (range 0–7.1%). There was good agreement (kappa = 0.65) between the measurement on the left and the right sides at all ages. Of the investigated connective tissue signs, thin transparent skin was noted in 0.1%, blue sclerae in 0.1%, and an elevated palate in 2.3% of the children. It was observed that 8.2% of the children were able to touch their nose with their tongue (Gorlin's sign) and 23.7% were able to touch their chin. The other signs were not observed in any of the children.

Conclusion. We validated the criteria for performing the Beighton Score in (Dutch) children. A cutoff point of ≥ 5 should be chosen for Dutch children aged between 4 and 9 years. It appeared that one-sided Beighton Score could be determined during screening of healthy populations. Additional diagnostic tests are warranted if one of the following signs, such as a thin transparent skin, hyperelastic skin, large hematomas, prominent scars, molluscoid tumors, striae distensae, blue sclerae, elevated palate, and Gorlin's sign, is encountered in a child. Although it is not possible to extrapolate the data, we expect that the findings are relevant for children of Caucasian race. (J Rheumatol 2001;28:2726–30)

Key Indexing Terms:

JOINT HYPERMOBILITY | CONNECTIVE TISSUE SIGN | BEIGHTON SCORE
PREVALENCE | VALIDATION | CHILDREN

One may consider a connective tissue disorder such as Ehlers-Danlos syndrome (EDS), benign joint hypermobility syndrome (EDS type III), or Marfan syndrome if a child presents with signs such as joint hypermobility, hyperelastic skin, and is suspected of connective tissue disease, etc. Special attention must be paid to inspection of the skin, the eyes, the vascular systems, and the mobility examination[1].

For quantitation of connective tissue signs such as hyperelastic skin, hematomas, scars, and joint hypermobility that indicate a connective tissue disorder, a scoring system is available only for joint hypermobility.

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Several instruments are available for scoring joint mobility[2-6]. The most widely used is the Beighton hypermobility score[2], because it can be used easily and is suitable for epidemiological studies[7]. Such a study was first reported in 1964 by Carter and Wilkinson[6] and it was modified in 1972 by Beighton[2]. The investigations described by Beighton consist of observing the results of movements, whereby 9 measurements are scored. One point is scored if the result of a movement is beyond the standard. The minimum achievable score is 0; the maximum is 9[2-4]. Total score of 4 is generally used as a cutoff point[4,5,8,9]. According to Beighton, one may speak of generalized hypermobility if the score is ≥ 4. The influence of sex[2-4,7,10-15], age[2,4,7,10,12,13,15,16], and race[2,4,10,12] on mobility have been extensively reported. In general, it can be stated that women are more mobile than men, that mobility declines with advancing age, and that Asians are more flexible than Caucasians. There is also a relationship between dominant side and hypermobility. Generally, a lower score is achieved on the dominant side than the nondominant side. Increased muscle tension has been reported as a possible cause of decreased mobility on the dominant side[2,4,9,10].

The prevalence of hypermobility has been reported to vary from 5% to 43%[4,14]. Such a large variation may be

explained by the use of different measuring instruments and different cutoff points in the use of the Beighton Score. The investigated groups also varied with regard to age and ethnic origin.

In 1997, Rikken-Bultman, *et al*[9] investigated the occurrence of hypermobility in children in The Netherlands. The prevalence of hypermobility at a cutoff point of ≥ 4 in children aged 4–12 years was 15.5%. However, in that study the mobility of the whole age group was looked at with no recommendations on the use of the Beighton Score in Dutch children.

There are scarce reports on the prevalence of the other connective tissue signs. In 1995, Grahame and Pyeritz[17] reported that 7% of children with Marfan syndrome had striae, 22% had papyrus scars, and in 63% of the children one could speak of an increased skin elasticity[17]. In 1998, El-Garf, *et al*[10] investigated 997 healthy Egyptian children. An elevated palate was noted in 37% and there was a significant relationship with joint hypermobility.

Another characteristic that points to a connective tissue disorder is the ability to touch the nose with the tip of the tongue (Gorlin's sign). In the normal population only 10% are able to do so, whereas this is the case in 50% of patients with EDS[18,19]. Skin elasticity decreases with advancing age[20]. Grahame[21] reported that women had increased skin elasticity compared with that in men, whereas this difference in children was insignificant.

The occurrence of connective tissue signs has not been investigated extensively in healthy children. Our aim was (1) to validate the Beighton Score for Dutch children, and (2) to determine the prevalence of connective tissue signs fitting in with a particular connective tissue disorder.

MATERIALS AND METHODS

Investigators. The 2 investigators were physiotherapy students. The measurement instruments used by Beighton were taught to them by an experienced pediatric physiotherapist. They received one month's training on examining connective tissue disorders at the outpatient pediatric dermatology unit (A.P.O.).

Pilot study. Prior to collecting the particulars, a pilot study into the inter-investigator variation in the Beighton score was conducted. For this purpose, 48 children from a primary school in Capelle/IJssel were divided according to age (4–12 years) and sex, and were examined. Both investigators were unaware of each other's results. A score of 4 was chosen as a cutoff point for hypermobility, based on the figure reported in the literature[5,8-10].

Investigated population. Three schools in average size municipalities of Vlaardingen, Schiedam, and Poortugaal in the vicinity of Rotterdam were approached to participate in the study. All parents received written information after approval by the director and the school board of each school. The teachers provided the date of birth of each child, the eventual presence of a movement disorder, and the ethnic origin of the child if it was other than Dutch. Inclusion criteria were parental permission and age between 4 and 12 years. Exclusion criteria were illness or syndromes that are accompanied by structural changes in the connective tissue and pain in the joints.

A total of 960 parents were approached. Refusal to participate in the study was received from 127 parents. Twenty-seven parents did not respond. Eight children were ill on the day of examination. Three children dropped out because of a movement disorder including juvenile chronic arthritis.

A total of 795 children were examined. Six children were excluded because they were found to be older than 13 years. Ethnic origin of 16 children was non-Dutch. These 16 children were excluded from the study because hypermobility is race dependent and the non-Dutch group was too small to allow any objective conclusion. Thus a total of 773 children were included in the study.

Examination procedure. The examination was conducted in combination with a physical education lesson so that all the children were dressed in a T-shirt and shorts. The examination room had a pleasant temperature. The children were examined prior to participating in the physical education activity to avoid any eventual influence of movement on the mobility. All the children were familiarized with the examination procedure using photographs. The children were asked to raise their T-shirt in order to score several of the items on the connective tissue sign examination list.

Besides scoring the hypermobility and the connective tissue signs, dominant hand was also noted. For the latter purpose, young children were asked to draw something. Older children were asked to indicate the hand used for writing.

Beighton hypermobility score. Beighton hypermobility score was chosen to measure joint mobility because this method is used in many studies[7], and as well the examination could be conducted easily and required little time[22]. The measuring instrument of Beighton consisted of 5 items (Table 1). The first 4 items are conducted both on the right and the left sides. Each item in which one could speak of hypermobility scored 1 point. The total score was a minimum of 0 and a maximum of 9 points.

The child did the active extension of the knee and elbow. The investigator was allowed to lead the movement, but not conduct it passively. The investigator conducted the other items. The degree of the movement was estimated. The examination caused no pain.

Examination into connective tissue signs. Examination into abnormal connective tissue signs concerned skin elasticity, presence of hematomas, scars, molluscoid tumors, striae distensae, blue sclerae, position of the palate, and joint mobility. The mobility of the tongue was also examined. A description of the examination items is given in Table 2[23].

Statistical analysis. The chi-square test was used to test differences in the distribution of a categorical variable between 2 groups. The Spearman rank correlation coefficient test was used to test a monotonous relationship between 2 ordinal variables. The Wilcoxon rank-sum test was used to test differences between 2 paired observations of an ordinal variable. The McNemar test was used to test differences between 2 paired measurements of a dichotomous variable. The kappa coefficient was used to test the level of agreement between 2 paired measurements of the same score.

RESULTS

A total of 773 children (395 boys, 378 girls) were included in this study. The differences in the prevalence of both hypermobility and the connective tissue signs between the boys and the girls were not significant (p values ranging from 0.115 to 1.000), except for the sign of the ability to touch the chin with the tongue ($p = 0.005$). In particular, 77 boys and 106 girls (respectively 19.5% and 28% of the total investigated group) could touch their chin with their tongue.

Pilot study. Good inter-investigator agreement for the total Beighton score (kappa = 0.81) was observed in the pilot study.

Joint mobility. A score ≥ 4 was observed in 20.8% (range 0–49%) of the children. There was a significant negative rank correlation ($r_s = –0.451$, $p \leq 0.0005$) when one looked

Table 1. The Beighton score.

Activity	Points (0-9)
Passive dorsal flexion of the little finger of more than 90° with wrist in the mid position	1 point for each little finger
Passive movement of the thumb, so that the thumb touches the ventral side of the lower arm	1 point for each thumb
Active extension of the elbow of more than 10°	1 point for each elbow
Active extension of the knee of more than 10° lying on the back	1 point for each knee
Bending forward with stretched knees so the palms touch the ground	1 point

Table 2. Description of important examination items in Ehlers-Danlos or Marfan and related syndromes.

Symptom	Description
Thin transparent skin	Is the vessel plexus directly visible and does the skin give an impression of aging incompatible with the calendar age?
Hyperelasticity	How far can the skin be stretched in the elbow position of 90°? How far can the skin of the neck be stretched laterally?
Hematomas	Are there large prominent hematomas?
Scars	Are there large and atrophic scars visible?
Molluscoid tumors	Are there encapsulated fat "herniations"?
Striae distensae	Are there long, stretched, somewhat sunken, wrinkled skin patches?
Blue sclerae	Is there any indication of blue coloring in the white of the eye?
Palate	What is the position of the palate?
Mobility of the tongue	Can the tip of the tongue touch the nose (Gorlin's sign) and/ or the chin?

at the Beighton score according to age: the older the child, the lower the score. Generalized hypermobility was observed in 5.8% of the Dutch children aged 4–9 years when a cutoff point ≥ 5 was used. Generalized hypermobility was observed in 5.3% of children older than 10 years using a cutoff point ≥ 4 (Figure 1).

There were 644 (83.3%) right-handed children, 123 (15.9%) left-handed children, and 6 (0.8%) children had no dominant hand. There was a good degree of agreement (kappa = 0.65) between the measurements on the left and the right sides. It appeared that when there was a discrepancy in the measurements of both the sides, the nondominant hand (n = 109) did score significantly higher than the dominant hand (n = 68) (p = 0.001).

Connective tissue signs. The investigated connective tissue signs are shown in Table 3. Prevalence is given per sign. It can be seen that most of the connective tissue signs occurred only incidentally; the exception was the sign of the ability to touch the chin with the tongue in 23.7% of the children. No sign showed a significant correlation with the age (Table 4).

DISCUSSION

In our investigations into joint hypermobility using the Beighton score with a cutoff point of ≥ 4, a "generalized hypermobility" would be observed in 20.8% of the children. The impression of Beighton is that the majority of musculoskeletal complaints attributable to hypermobility occur in the most supple 5% to 10% of the population[4]. However, children with musculoskeletal complaints were excluded from our study. If we had included these children, we expect that we would find 25% to 30% "generalized hypermobility" with the cutoff point of ≥ 4. There is no reason to assume that there is a higher percentage of hypermobile children in The Netherlands. Thus, in our opinion, the cutoff point should be readjusted to ≥ 5. Generalized hypermobility was observed in 5.8% of Dutch children aged 4–9 years when a cutoff point of ≥ 5 was used. A cutoff point ≥ 4 must be used from the age of 10 years; generalized hypermobility is observed in 5.3% of these children.

We did not investigate these hypermobile children according to the criteria of benign joint hypermobility syndrome (BJHS) as described by Graham, *et al*[24]. So we

Table 3. ^Prevalence of connective tissue characteristics in 773 children.

Symptoms	Prevalence, n / %
Thin transparent skin	1 / 0.1%
Hyperelasticity	—
Hematomas	—
Scars	—
Molluscoid tumors	—
Striae distensae	—
Blue sclerae	1 / 0.1%
Palate	18 / 2.3%
Gorlin's sign (nose)	63 / 8.2%
Tongue to chin	183 / 23.7%

Table 4. Correlation between connective tissue symptoms and age.

Symptom	p	Correlation Coefficient
Thin transparent skin	0.561	–0.21
Blue sclerae	0.212	–0.45
Palate	0.079	0.63
Mobility of the tongue	0.688	–0.14

cannot give an indication of the prevalence of BJHS in the investigated Dutch population.

Joint mobility depends on the ethnic background. One can assume that children with the same ethnic background would have the same percentages of hypermobility. This could not be confirmed compared with other studies[6,7,10,11,13,16]. An important reason for this is the differences in the measuring instruments, differences in the age groups, and the use of the Beighton score with different cutoff points in these studies.

Our results are comparable with those reported by Rikken-Bultman, *et al*[9], who observed generalized hypermobility in 15.5% of 252 children. Thus, at a cutoff point of ≥ 4, there is a difference of 5% with the present study in which 20.8% of the children were scored as having generalized hypermobility. A possible explanation for this discrepancy cannot be given.

In this study, it is noteworthy that there is a negative correlation between mobility and age; the older the patient, the more reduced is the mobility. Similar observations were reported in other studies[2,7,10,13,15,16]. No correlation was observed between age and any of the other connective tissue signs.

There was good agreement between the measurements on the left and right sides. If the measurements were discrepant, the nondominant side scored the highest, corroborating results in other studies[2,4,9]. Thus, it is sufficient to conduct only a one-sided Beighton score examination when screening healthy populations. The duration of the hypermobility examination could thus be shortened by about 2 minutes[22]. In calculating the total score one must multiply the score of extremity by 2. If one uses the Beighton score for a scientific study, then one must conduct 2-sided measurements — the Beighton score is conducted in this manner worldwide.

The difference between the boys and the girls is not significant for any connective tissue signs fitting in with a connective tissue disorder. As well, no difference between boys and girls of the same age group was reported in several other studies[9,10,14,21]. A significant difference between the boys and girls who were older than 12 years was reported[2,7,14,21].

At the beginning of this study, it was expected that there might be a correlation between the various connective tissue signs. However, not a single significant correlation was found. Further, it appeared that with regard to the connective tissue signs, there was only one examination item that was regularly encountered in the children. This was the ability to touch the chin with the tongue in 23.7% of the children.

A thin, transparent skin, hyperelastic skin, large prominent hematomas, scars, molluscoid tumors, striae distensae, blue sclerae, an elevated palate and Gorlin's sign were either absent or were encountered to only a limited extent (8.2%). These items may be of discriminatory value at physical examination.

The incidence (8.2%) of Gorlin's sign agrees well with the incidence (10%) reported in other studies[18,19]. The incidence of an elevated palate was reported to be 37% by El-Garf, *et al*[10]. This figure exceeds our observation by about 35%. A possible explanation for this discrepancy may be that establishing an elevated palate is subjective.

We have tried to validate the Beighton criteria for our population of Dutch children. According to our data, we are of the opinion that from 4 to 9 years of age a Beighton score with a cutoff point of ≥ 5 is suitable and above 10 years of age, a cutoff point ≥ 4 must be used. As well, we concluded that the Beighton criteria could be applied one-sided in a healthy pediatric population. Since the particulars on the prevalence of connective tissue signs fitting in with a connective tissue disorder have now been compiled, additional investigations into the occurrence of these signs in children with a diagnosed connective tissue disorder are recommended.

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