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Original article

Gleno-humeral abduction measurement in patients with Ehlers-Danlos syndrome

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ABSTRACT

Background: No genetic tests or other investigations are available to establish the definitive diagnosis of Ehlers-Danlos syndrome (EDS). A presumptive diagnosis can be made based on a converging set of findings, including a family history of the disease, the patient's medical history, and the physical findings. The Beighton score is currently the reference standard tool for assessing joint hypermobility, which must be present in at least 5 of 9 tested joints. However, joint hypermobility testing may be challenging, for instance in patients with pain during mobilisation and/or tight hamstring muscles. Furthermore, the Beighton score may be less than 5 in patients with other unequivocal evidence of EDS. The objective of this study was to assess the contribution of gleno-humeral abduction measurement to the diagnosis of EDS.
Hypothesis: Gleno-humeral abduction measurement using a standardised method assists in the diagnosis of EDS.
Methods and material: Retrospective case-control study comparing 110 patients with known EDS (cases) to 100 controls.
Results: Gleno-humeral abduction was significantly greater in the cases than in the controls, irrespective of age, sex, and Beighton score. Gleno-humeral abduction beyond 90° was 92.5% sensitive and 96.4% specific. Inter-examiner reproducibility of gleno-humeral abduction measurement was excellent.
Discussion: Increased gleno-humeral abduction may be sufficient to demonstrate joint hypermobility and to suggest EDS in patients whose personal and family history is consistent with this diagnosis.
Level of evidence: III, case-control study.

1. Introduction

The diagnosis of Ehlers-Danlos syndrome (EDS) currently rests on a converging set of arguments, most of which are obtained from the medical interview. No conclusive genetic testing strategy exists to date and, despite promising results reported with diffusion-weighted magnetic resonance imaging, no available investigations can provide the definitive diagnosis. Consequently, testing a clinical suspicion of EDS must rely on the physical examination [1–3].

Physical findings suggestive of EDS that can be identified by simple observation include abnormal gait and/or posture, bruising, stretch marks, and atrophic scars. The main footprint abnormality is a flat or even rounded forefoot. Hyperextensibility and a velvety texture of the skin should be sought by palpation [4]. However, examination of the joints with determination of the Beighton hypermobility score [5] is often the most informative part of the physical examination. Hypermobility manifests most often at the thoraco-lumbar spine during forward flexion, during flexion of the thumb to the forearm, during dorsiflexion of the fifth metacarpo-phalangeal joint, and as hyperextension of the elbows and knees. Recording joint-mobility at each of these joints on both sides provides the Beighton score.

However, joint-mobility in a given patient varies over time depending on pain severity on the day of testing, patient cooperation, and age-related joint stiffening. A reliable, easy-to-use, reproducible joint-mobility parameter measurable using a goniometer and unlikely to be affected by age would therefore be useful. Gleno-humeral mobility might be such a parameter.

The objective of this study was to assess the contribution of gleno-humeral abduction measurement to the diagnosis of

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Fig. 1. Position for measuring the range of gleno-humeral abduction.

EDS. The working hypothesis was that gleno-humeral abduction measurement using a standardised method would assist in the diagnosis of EDS.

2. Material and methods

2.1. Study design and participants

A retrospective study was performed to compare patients with (cases) and without (controls) EDS. The cases were seen at an EDS clinic located in the suburbs of Orleans, France, for any of the following reasons:

- referral for specialised management after a definitive diagnosis of EDS was made elsewhere;
- referral by the Hôtel-Dieu physical medicine and rehabilitation department (Prof. Hamonet) in Paris (France) for a strong suspicion of EDS, for convenience related to place of residence or long waiting times at the Paris site;
- patient concern about possibly having EDS after searching the internet and social media for relevant information;
- EDS diagnosed by the author after more or less lengthy diagnostic wanderings;
- or referral because of known EDS in one or more relatives.

None of the patients was referred by specialists of mechanical shoulder disorders. All cases included in the study met medical-interview criteria for suspected EDS [4].

The controls were selected at random among the author's patients. Exclusion criteria were suspected or known EDS, adhesive capsulitis of the shoulder, inflammatory joint disease, a history of shoulder injury, and cervico-scapular pain.

2.2. Assessment method

Gleno-humeral abduction was measured in all cases and controls by the author using the following standardised technique (Fig. 1). The patient is naked to the waist and stands in front and to one side of the examiner. For the left shoulder, the examiner places the right thumb and forefinger on either side of the tip of the scapula. The examiner's left forearm supports the patient's left arm, which is in neutral rotation with the elbow flexed. The examiner then passively abducts the patient's shoulder, until the scapula just starts to tilt. At that point, the angle formed by the patient's left arm and the vertical is measured using the fixed arm of the goniometer. The angle is then recorded on the other side [6].

Gleno-humeral abduction measurement is usually painless. Some patients experience pain beyond a certain range of abduction and the measurement test must then be stopped. However, in this study pain never arose within the normal range of abduction (90°). The test may also have to be stopped if the examiner deems that further abduction might cause dislocation of the gleno-humeral joint.

2.3. Reproducibility assessment

Inter-examiner reproducibility was assessed by having five students pursuing a degree in EDS examine both shoulders of 2 cases. The mean value obtained by the 5 students was then compared to the mean value obtained by the author.

3. Results

3.1. Study participants

The study included 110 cases. Mean age was 30 years (range, 6–69 years) overall, 32.5 years (range, 6–66 years) in the 87 females, and 22 years (range, 6–69 years) in the 23 males.

In addition, 100 controls were studied. Mean age was 48 years (range, 12–87 years) overall, 50 years (range, 23–83 years) in the 50 females, and 45 years (range, 12–87 years) in the 50 males.

3.2. Gleno-humeral mobility in the overall group of cases

The 110 cases had a mean Beighton score of 5 (range, 0–9). Only 4 cases had a normal range of gleno-humeral abduction of 90°; the other 106 had higher values with a mean of 116° (range, 95°–150°) (Fig. 2). Thus, the mean value in the cases was 29% higher than the normal value. Gleno-humeral joint hypermobility was 96.4% sensitive for EDS in the group of cases.

Of the 220 gleno-humeral mobility measurements performed in the cases, only 1 resulted in a complication, which consisted in subluxation of the gleno-humeral joint. Reduction was performed immediately by the examiner. No residual abnormalities were evidenced during the follow-up evaluation 4 months later.

3.3. Gleno-humeral mobility in subgroups of cases

The patients were divided into subgroups by sex, age (6–40 years vs. >40 years), and Beighton score. The mean Beighton score was not significantly different between the females (5.22) and the males (4.96). Mean gleno-humeral abduction was also similar in the females (116°) and males (117.7°). Mean gleno-humeral abduction was 118.6° in the 77 cases aged 6 to 40 years and 110.6° in the 33 cases older than 40 years (Table 1). Mean Beighton scores in these two age groups were 6.6 and 4.55, respectively (Table 2). Table 3 reports the Beighton scores according to the range of gleno-humeral abduction. Of the 4 cases with normal gleno-humeral abduction (90°), 3 were females with Beighton scores of 2, 3, and 7, respectively; the male had a Beighton score of 2.

3.4. Gleno-humeral mobility in the controls

In the control group, gleno-humeral abduction was greater than 90° in 7 of the 100 shoulders in females (95°, $n = 3$; 100°, $n = 2$; and 130°, $n = 22$) and in 8 of the 100 shoulders in males (95°, $n = 7$; 100°, $n = 1$). Thus, a total of 15 of 200 control shoulders had an excessive range of gleno-humeral abduction, yielding a specificity of 92.5%.

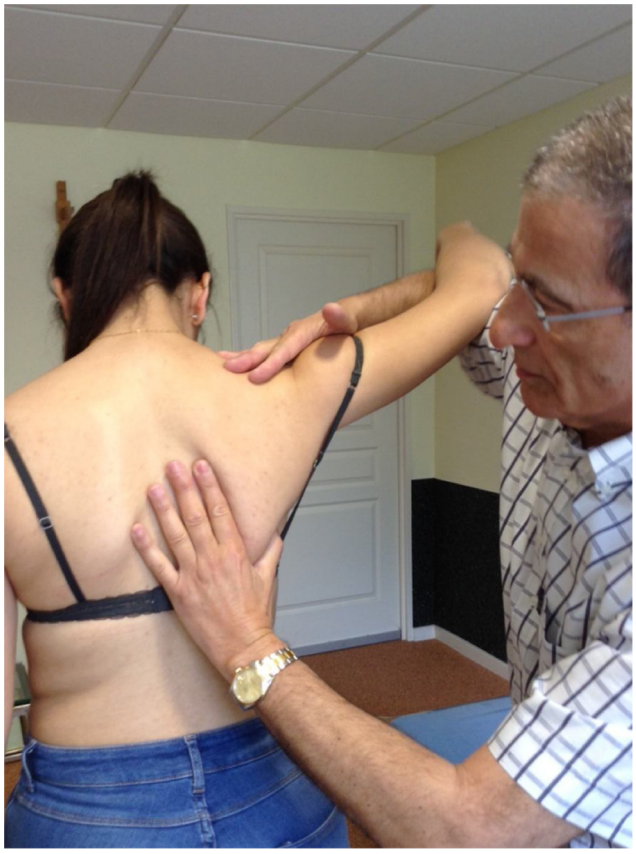

Fig. 2. Abduction range is 110°, with no outwards scapular tilting over the rib cage.

Table 1
Mean gleno-humeral abduction by age group.

Age groups, (years)	Females, (*n*)	Males, (*n*)	Abductionn, (°)
6–10	6	6	115
10–20	16	8	120
20–30	15	4	121
30–40	20	2	117
40–50	18	2	113
50–60	10		111
>60	2	1	108

Table 2
Mean Beighton score by age group.

Age groups, (years)	Number of patients	Mean Beighton score
6–10	12	6.5
10–20	24	5.35
20–30	19	5.95
30–40	22	5.06
40–50	20	5.22
50–60	10	3
>60	3	5.33

Table 3
Mean gleno-humeral abduction at each Beighton score value.

Beighton score	Number of patients	Gleno-humeral abduction, (°)
0	7	110
1	1	116
2	13	108
3	7	106
4	17	115
5	7	120
6	22	117
7	14	119
8	12	125
9	10	121

Table 4
Inter-examiner reproducibility of the passive abduction test.

	1st patient	1st patient	2nd patient	2nd patient
Side	Left, (°)	Right, (°)	Left, (°)	Right, (°)
Author	90	95	100	100
Student 1	90	90	93	95
Student 2	90	90	100	100
Student 3	85	90	105	110
Student 4	90	90	95	110
Student 5	92	92	95	95

3.5. Reproducibility of gleno-humeral abduction measurement

As shown in Table 4, mean gleno-humeral abduction of the 4 shoulders used for reproducibility testing was 96.25° (range, 90°–100°) when measured by the author and 94.85° (85°–110°) when measured by the five students. The 1.40° difference is smaller than the standard deviation of 1.9°, indicating good inter-examiner reproducibility.

4. Discussion

The ability to place both palms flat on the floor when bending forward with the knees extended is a typical sign of EDS. It may be difficult to assess, however, for instance if the patient is wheelchair-bound, experiences the low back pain common in EDS, and/or has tight hamstrings [4]. The demonstration of joint hypermobility may be hindered if pain caused by passive joint mobilisation leads the examiner to stop the manoeuvre. Thus, pain may arise in the multiple joints involved in touching the tip of the thumb to the forearm [7], even when the test is performed by the patient. Similarly, joint pain may be an obstacle to determining whether the fifth metacarpo-phalangeal joint can be dorsiflexed beyond 90° or whether the elbows and/or knees are hyperextensible. In addition, testing the elbows and knees for hyperexensibility involves grasping the limb on either side of the joint, which may also cause pain due to the cutaneous hyperesthesia seen in EDS.

Shoulder hypermobility is usually defined as excessive external rotation [8]. However, in a patient at risk for multidirectional shoulder instability, testing for excessive external rotation by passive mobilisation may cause dislocation. Gleno-humeral abduction may therefore be a better criterion. Gleno-humeral abduction contributes to lateral elevation of the arm, which involves three phases [9] (Fig. 3). The first phase, from 0° to 90° occurs within the gleno-humeral joint and stops when the lesser tuberosity of the humerus comes into contact with the upper rim of the glenoid. In the second phase, from 90° to 150°, the gleno-humeral joint is blocked at 90°

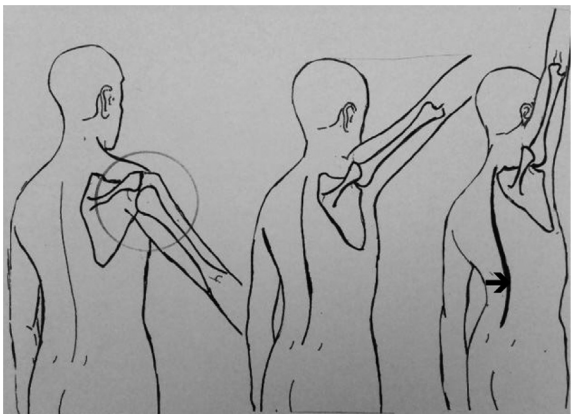

Fig. 3. The three stages of lateral elevation of the arm.
Kapandji I. A. Physiologie articulaire, tome 1 : membre supérieur.

and further elevation occurs due to the scapula tilting outwards. Scapular tilt can elevate the arm to about 150°. Finally, the third phase raises the arm laterally from 150° to 180° via lateral bending of the spine to the contralateral side. Outwards tilting of the scapula is easily perceived before 90° of abduction and becomes marked beyond this range [10]. In patients with adhesive capsulitis, outwards scapular tilting occurs well before 90° of abduction [11].

Beighton score values may be highest during childhood [12]. Children are generally free of low back pain, which can prevent the patient from placing the palms flat on the floor when bending forwards with the knees extended, thereby decreasing the Beighton score by 1 point. In addition, hamstring tightness related to a growth velocity mismatch between the soft tissues and the distal femurs and proximal tibias usually develops around puberty.

Our comparison of EDS patients younger and older than 40 years showed that mean gleno-humeral abduction was 8° less in the older patients, despite remaining excessive. Beighton pointed out that joint stiffness developed with advancing age in the joints used to determine this score [13]. However, gleno-humeral abduction was not restricted in our control group, suggesting that the gleno-humeral joint may be spared by age-related joint stiffness.

In the Brighton criteria set for joint hypermobility syndrome [14], one of the two major criteria is a Beighton score ≥4 and one of the eight minor criteria is a Beighton score of 1, 2, or 3; or of 0, 1, 2, or 3 if age is over 50 years. It is thus worth noting that in our study 96.4% of patients with EDS had excessive gleno-humeral abduction, irrespective of age and sex. Thus, examining the gleno-humeral joint nearly consistently demonstrates hypermobility in patients with EDS.

5. Conclusion

This study shows that, in patients with a strong suspicion of EDS based on symptoms, the family history, and the physical findings [15], a Beighton score below 5 does not rule out EDS. In the 45 patients whose Beighton score was 0 to 4, mean gleno-humeral abduction was 20° higher than the normal value. In the 65 patients whose Beighton score was 5 or higher, the mean excess abduction was 30°. Thus, a larger number of hypermobile joints was associated with greater gleno-humeral hypermobility.

The 96.4% sensitivity and 92.5% specificity of gleno-humeral hypermobility indicate good diagnostic performance, particularly given the good inter-examiner reproducibility. In patients whose medical history suggests EDS, gleno-humeral abduction measurement may thus provide proof of joint hypermobility, even when the Beighton score is less than 5, as was the case in 45 (41%) of our patients.

Disclosure of interest

The author declares that he has no competing interest.

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None.

Contributions

Not applicable.

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