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Clinical Assessment Methods for Classifying Generalized Joint Hypermobility

FOR NON-EXPERTS

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There is lack of knowledge about which clinical assessment methods are best suited for classifying Generalized Joint Hypermobility (GJH), here we review them systematically. Four test assessment methods were inspected (Beighton Score [BS], Carter and Wilkinson, Hospital del Mar, Rotes-Querol) and two questionnaire assessment methods (Five-part questionnaire [5PQ], Beighton Score-self reported [BS-self]). Studies were rated "fair" or "poor." The recommendation for clinical use in adults is BS with cut-off point of 5 of 9 including historical information, while in children it is BS with a cut-off point of at least 6 of 9. However, more studies are needed before evidence-based recommendations can be made.

Introduction

Generalized joint hypermobility (GJH) is relatively common, occurring in about 2–57% of different populations. Important reasons for this may be the use of many different clinical assessment methods and criteria for classification. GJH is characterized by joints moveable beyond the normal range of motion in multiple joints. Many people with GJH do not have symptoms, which also makes it difficult to accurately estimate the number of people with this condition. When GJH is accompanied with symptoms, it is defined as a health-related disorder, for example, Joint Hypermobility Syndrome (JHS) or the Ehlers-Danlos Syndrome – Hypermobile Type (hEDS). The two conditions (JHS and hEDS) have very close overlap to the point of being indistinguishable; here it is referred to as JHS/hEDS. The condition of JHS/hEDS can be defined as a connective tissue disorder, characterized generally by GJH, complications of joint instability, pain, characteristic skin signs, and reduced quality-of-life. Until now, JHS has been diagnosed by the Brighton tests and criteria, and hEDS by the Villefranche criteria, both including the Beighton scoring (BS) system of nine tests for assessment of GJH.

In particular two measurement properties are evaluated in the clinical assessment methods: Validity and reliability. Validity refers to whether the result can be confirmed as correct, while reliability refers to whether a result can be repeated with the same or similar result.

BS consists of one test of the low back and lower extremities, and four bilateral tests elsewhere (first finger opposition, fifth finger extension, elbow extension, knee extension, and back forward bending), with scores ranging from 0 to 9. Influencing factors on BS are age, gender, ethnicity, and physical fitness. For adults, a cut-off point of 4/9 for GJH was included in the Brighton criteria for JHS, while 5/9 for GJH is the criteria for hEDS in the Villefranche criteria. For children, there was no consensus on a specific cut-off point for GJH, but 5/9, 6/9, and 7/9 have been suggested. Reliability of Beighton or similar tests is known to be good, but there is lack of evidence for the validity of this method.

The five-part questionnaire (5PQ), so far used only for adults, consists of five questions, including actual and historical information about generalized joint hypermobility (forward bending of the back, first finger opposition, the ability to amuse friends with strange body shapes, dislocation of shoulder/knee, perception of being double-jointed). The 5PQ is generally considered reliable but would benefit from more investigation on several kinds of validity. Clear diagnostic clinical assessment

methods for GJH with and without symptoms are needed, both for diagnosing JHS/hEDS and measuring treatment effects of JHS/hEDS, in children as well as in adults.

In summary, there is lack of knowledge of which clinical assessment methods are suitable for classifying GJH. Therefore, the purpose of this study was to perform a systematic review identifying the clinical assessment methods for classifying GJH, to evaluate their reliability and validity according to the recommended COSMIN procedure, and finally to summarize the best evidence.

Results and Discussion

Four test assessment methods (BS, CW, HdM, RQ) and two questionnaire assessment methods (5PQ, BS-self) were identified in the literature for classifying GJH, in children and adults. Most studies were on BS, and only BS and 5PQ reported aspects of validity. The majority of the reliability studies showed some positive evidence for BS, and thus, may seem acceptable to be used in clinical practice, provided that testing is done in the same way everywhere. There are shortcomings on studies for the validity of BS, while the three other test assessment methods (CW, HdM, RQ) lack enough information on both reliability and validity to recommend.

For the questionnaire assessment methods, most of the studies were reported on 5PQ, which shows to be a promising assessment method for future population studies. The additional questionnaire, BS-self, may also seem promising, as it contains illustrations of the test procedures for each of the BS tests. However, the questionnaire assessment methods need more evaluation before they can be used clinically, since very few studies have reported measurement properties on their reliability and validity.

Since JHS and hEDS in the current review are recognized as one and the same condition, a specific cut-off point needs to be decided, and 5/9 may be suggested for future use in adults. However, since joint mobility, and therefore, BS is known to decrease by age, there is a need for adults also to include additional historical information. Since children have individual growth periods, this may be the reason for using two cut-off points (a lower and an upper) taking age and gender into consideration. The upper cut-off point is suggested to be at least 6/9 as used in previous studies. Warming up before performing flexibility tests may influence the outcome of a test assessment method. However, almost no studies reported whether participants did warm up, and the influence of such performance is therefore, unknown.

This review highlights a number of areas needing future research. Because of the limited studies on the clinical assessment methods for classifying GJH, more high-quality studies, especially those evaluating different aspects of validity, are required. Additional test assessment methods other than the presently described may be considered in order to support the presence of GJH in the diagnostic procedure of heritable connective tissue disorders. Also of importance is that consensus is warranted regarding selection of specific test and questionnaire assessment methods for classifying GJH, the test performance, and the cut-off points by which age, gender, and ethnicity may be taken into account. Limitations of this review are the small number of studies, for which reason it was decided only to rate reliability and few types of the validity aspect. Further, since the sample size in clinical studies is often much smaller than in questionnaire studies, it is recommended that minor sample sizes using test assessment methods should not be rated as strictly as when using questionnaire assessment methods. In spite of these limitations, this review benefits from being systematic and applying recommended evaluation strategies for this kind of review.

Conclusion

In the current systematic literature review, four test and two questionnaire assessment methods for classifying GJH were found. Most of the studies used the Beighton Score. The reliability of this method seems acceptable. Shortcomings were found in studies on the validity of BS, while the three other test assessment methods lack information on both reliability and validity. Regarding questionnaire assessment methods, 5PQ is used most often, but only in adults. In conclusion, provided the testing procedures are consistent, the recommendation for clinical use in adults is BS with a cut-off point of 5 out of 9, including historical information. In children it is BS with cut-off point of at least 6 out of 9. Although the Beighton Score is recommended for classifying GJH, more studies are needed, especially on the validity properties of these assessment methods.

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