Postural Sway and General Joint Hypermobility

Introduction

Individuals with joint hypermobility, also known as generalized joint hypermobility (GJH), experience synovial joint movement "beyond the normal limits of the range of motion, taking into consideration the age, gender and ethnic background in otherwise healthy subjects".[1] Some individuals with GJH report no pain or difficulties participating in daily and recreational activities, while others experience pain, dislocation or subluxation, inflammation, or various other symptoms in the hypermobile joint in the absence of systemic causes. Individuals experiencing these symptoms are classified as having Benign Joint Hypermobility Syndrome (BJHS)[2] or Joint Hypermobility Syndrome.[4] Heritable disorders of connective tissue can result in joint hypermobility,[3] such as Ehleros-Danlos, Marfan syndrome, and osteogenesis imperfecta; however, this review will focus on GJH and BJHS.

Various methods are available to assess GJH; the Beighton and Horan Joint Mobility Index (BHJMI) is the most frequently utilized measure (Figure 1 a-e). This tool evaluates an individual's ability to complete nine maneuvers: passively extend each little finger beyond 90 degrees, oppose each thumb to the forearm, hyperextend each elbow beyond 10 degrees, hyperextend each knee beyond 10 degrees, and forward flex the trunk to place the hands flat on the ground.[6] A point is given for each maneuver achieved, and a total score is summed, ranging from 0-9. Although no set standard cutoff score exists, a score of $\geq$4/9 is often used to classify GJH.[1]

To assess the presence of BJHS, the Brighton criteria is utilized (Figure 2).[8] Major and minor criteria are considered for BJHS diagnosis, and BJHS is defined as either the presence of two major criteria, one major and two minor criteria, four minor criteria, or two minor criteria and affected first-degree relative family history.[8] The two major criteria are a BHJMI score of $\geq$4/9 and the presence of arthralgia in four or more joints for longer than three months. The eight minor criteria include: 1) BHJMI scores 1-3 out of 9 (0/9 acceptable for adults 50+ years old), 2) arthralgia in 1-3 joints or back pain for longer than 3 months, 3) joint dislocations or subluxations in more than one joint or more than once in the same joint, 4) soft tissue inflammation and pain (e.g. epicondylitis, tenosynovitis), 5) Marfan habitus, 6) skin abnormalities (e.g. hyperextensibility of skin), 7) eye anomalies (e.g. drooping eyelids), and 8) varicose veins, hernia, or uterine/rectal prolapse.[8]

GJH occurs more frequently in females than in males[1,4] and is more often seen in younger than older individuals.[4] The prevalence of joint hypermobility may differ by race. Two reports[1,4] suggest that African and Asian populations are more likely than Caucasians to present with joint hypermobility. However, other studies suggest that Caucasians may be more likely to have joint hypermobility than African Americans.[22,23,24] Dissimilarities in the prevalence of GJH vary by population and differing definitions for GJH. The frequency of GJH ranges from 4.9% to 23.9% of the US population.[5]

The extent to which GJH affects physical function and balance is not well known. Previous studies have determined that adolescents to middle age adults with joint hypermobility have diminished joint proprioception,[7] which may diminish balance and contribute to a greater likelihood of injury or falls. Good static balance is critical for many basic functional activities, and assessment of postural sway stresses visual, vestibular and somatosensory-dependent functions of static balance, providing insights into the mechanisms driving balance impairments. The purpose of this review was to examine and summarize the current literature of studies that assesses the association between static balance, specifically postural sway, and joint hypermobility.

Methods

PubMed, CINAHL and Cochran databases were searched for articles pertaining to balance in the population of those with joint hypermobility (Figure 3). The terms searched included hypermobility, hypermobile, hyperlaxity, postural balance, one-leg stance, single-leg stance, postural sway, and static balance. Results were limited to those without Ehlers Danlos Syndrome and to those published in English.

A total of 28 articles were identified by the electronic search and were evaluated for relevance (Figure 3). Articles were included in the final selection if postural sway was measured during static balance activities. Articles were excluded if the focus of the study was balance interventions or the outcomes measured did not include postural sway. Articles were also excluded if the population of study participants was specific to those with a particular co-morbid condition (e.g. individuals with seizures, low back pain, or low postural tone). The remaining five articles are included in the literature review.

Results

Five observational, cross-sectional studies were reviewed (Table 1), which included a total of 300 participants. Three studies focused on participants between the ages of 20-30 years [9,10,12] while the remaining studies focused on participants with the average age of 14 years old.[11,13] Four of the five studies included only female participants.[9,10,12,13]

The BHJMI cutoff score for hypermobility varied between studies. The study by Iatridou et al.[9] classified hypermobility as a score of >4/9 while Mebes et al.[10] and Juul-Kristensen et al.[13] used a score of ≥6/9. Schmidt et al.[11] utilized multiple cutoffs: participants with BHJMI ≥4/9, ≥5/9, and ≥6/9. Aydin et al.[12] created sub-classifications of moderately hypermobile (BHJMI=3-4/9) and distinctly hypermobile (BHJMI≥5/9).

Several studies assessed overall postural sway, or center of pressure path length, while participants maintained various balance positions such as one-legged stance with eyes closed or double-legged stance with head extended and eyes closed. Studies by Iatridou et al.[9], Medes et al.[10] and Schmidt et al.[11] specifically assessed mediolateral and anteroposterior sway or deviation while participants maintained various positions.

Concerning mediolateral sway, Iatridou et al.[9], Medes et al.[10] and Schmidt et al.[11] reported increases in those with joint hypermobility during a one-legged stance with eyes open compared to participants without joint hypermobility. While Schmidt et al.[11] reported a similar finding for the subgroup with a BHJMI score of ≥4/9, there was no difference between those with BHJMI score ≥5/9 or ≥6/9 and the other participants. Schmidt et al.[11] reported increased mediolateral sway during single-legged stance with eyes open, single-legged stance with eyes closed and double-legged stance with eyes closed for those with BHJMI scores of ≥4/9 compared to other participants. Participants with a BHJMI score of ≥5/9 demonstrated increased mediolateral sway during single-legged with eyes closed, double-legged with eyes open and double-legged with eyes closed position.[11] Iatridou et al.[9] reported mediolateral sway increased in the one-legged stance with eyes open with head back position for participants with joint hypermobility.

According to Mebes et al.[10], there was no difference for single-legged stance with eyes open while Iatridou et al.[9] and Schmidt et al.[11] reported no statistically significant difference in mediolateral sway for those with joint hypermobility during a one-legged stance with eyes closed. This pertained only to the more hypermobile participants with BHJMI ≥5/9 in the study by Schmidt et al.[11] Schmidt et al.[11] further report no statistically significant difference for those with joint hypermobility during a double-legged stance with eyes open for those with the BHJMI score of ≥4/9 or ≥6/9 compared to other participants. Those with a BHJMI score of ≥5/9 had no difference in mediolateral sway for a single-legged stance with eyes open and for those with a score of ≥6/9 there also was no difference for single- or double-legged stance with eyes closed compared to those without joint hypermobility.

Regarding anteroposterior postural sway, Iatridou et al.[9] and Schmidt et al.[11] both reported an increase in sway in participants with joint hypermobility while maintaining various positions. Iatridou et al.[9] reported an increase in sway for those with joint hypermobility in the one-legged stance with eyes open and head extended position when compared to those without joint hypermobility. Schmidt et al.[11] reported increased sway in double-legged stance with eyes open for participants with ≥4/9 and ≥6/9 BHJMI scores and eyes closed in participants with ≥4/9 and ≥5/9 BHJMI scores. Participants with a BHJMI score of ≥5/9 also resulted in an increased anteroposterior sway in the one-legged stance with eyes closed for those with joint hypermobility.

None of the three studies[9,10,11] assessing anteroposterior sway reported statistically significant differences in the single-legged stance with eyes open for hypermobile participants. The study by Schmidt et al[11] also reported no differences for participants with a BHJMI score of ≥4/9 or ≥6/9 in the single-legged stance with eyes closed, those with a BHJMI score of ≥5/9 in a double-legged stance with eyes open or those with ≥6/9 score in the double-legged stance with eyes closed.

Overall sway assessments by Schmidt et al.[11] and Juul-Kristensen et al.[13] reported greater sway in those with joint hypermobility compared to those without for double-legged stance with eyes open and eyes closed. Schmidt et al.[11] further reported an increase in sway for all hypermobile participants in the single-legged stance with eyes closed and increase in overall sway for participants with ≥4-5/9 BHJMI score in the single-legged stance with eyes open. Aydin et al.[12] reported an increase in two positions for the distinctly hypermobile group; double-legged stance with head turned 45 degrees to the right on a firm surface with eyes closed and double-legged stance with head extended about 30 degrees while standing on a firm surface with eyes closed.

Schmidt et al.[11] and Juul-Kristensen et al.[13] both reported no statistically significance differences during the one-legged stance with eyes open for overall sway comparing the hypermobile to non-hypermobile participants. However, this was only true for the most hypermobile group in the Schmidt et al.[11] study with BHJMI scores ≥6/9. Aydin et al.[12] reported no significant increases in overall sway for the moderately hypermobile participants in all positions and no increases in sway for distinctly hypermobile participants in all positions aside from the two positions mentioned above.

Discussion

The aim of this literature review was to assess the association between joint hypermobility and postural sway. Postural sway can be measured by the change in center of pressure during stance positions, which reflects body movement to realign the center of body mass to maintain an upright position.[18] As previously mentioned, static balance requires visual, vestibular, and somatosensory input. Based on the articles included in this review, hypermobile participants demonstrated a significant increase in mediolateral sway when maintaining single-legged stance with eyes open. There was no consistent pattern for anteroposterior sway between participants with joint hypermobility compared to those without; however, this may be due to a lack of uniformity of the various positions held between studies and cutoffs for hypermobility. Regarding overall sway, several studies reported greater sway in hypermobile participants when maintaining a double-legged stance with eyes open and eyes closed. In general, participants with joint hypermobility had increased mediolateral, anterioposterior, and overall postural sway compared to those without joint hypermobility[9,10,11,13].

Several limitations were noted when comparing studies. First, the age of the participants varied from adolescents to young adults to those in their late twenties; no other age ranges were examined. Hypermobility and postural sway can vary by age. A study by Remvig et al.[14] noted that the prevalence of hypermobility is greatest in children and decreases in older adults. Tissue stiffening that occurs with aging may be one reason behind the reduced occurrence of joint hypermobility in older age groups.[1] A study by Rogind et al.[15] noted that postural sway increases as individuals age from 20 to 70 years of age. However, a study by Verbecque et al.[20] investigated the presence of postural sway in a natural bipedal stance for participants 2 to 18 years of age and reported a decrease in postural sway over time during these early years. Therefore, changes in joint hypermobility and postural sway based on age should be considered when comparing studies utilizing participants of different ages.

Second, the BHJMI score cutoff utilized in studies varied greatly. A BHJMI score of >4/9 is a frequently used cutoff for joint hypermobility. However, the cutoff score was inconsistent across the studies included in this literature review. The Schmidt et al.[11] and Aydin et al.[12] studies further divided participants with joint hypermobility into subgroups to assess outcomes based on a range of hypermobility. To account for increased hypermobility in younger participants, some studies[16,17,19] choose to increase the BHJMI cutoff points to 5/9, 6/9 or 7/9. The study by Juul-Kristnesen et al.[13] utilized this method when assessing participants with an average age of 14 years old. However, the study by Schmidt et al.[11] also included participants with an average age of 14 years but used 4/9 as the lowest cutoff for hypermobility. The study by Mebes et al.[10] involved participants with an average age of 28 years old, yet the cutoff score was higher at $\geq$6/9. The variation in the BHJMI cutoff score may alter associations of joint hypermobility and postural sway.

Third, the amount of stance time per position was variable across studies. Stance times per study included in this review ranged from 15 seconds to 60 seconds. It has been noted that holding a position for a longer duration may be more challenging and result in an increase in postural sway compared a shorter duration.[20] In addition, the number of trails also varied by study. The study by Iatridou et al.[9] assessed postural sway during one trial of 20-second stances for three different positions while the study by Schmidt et al.[11] assessed postural sway during two or three trials of 30-second stances for four different positions. Potentially, multiple trials with longer stance times could lead to fatigue which may in turn increase postural sway.[21]

Conclusion

In summary, mediolateral, anterioposterior, and overall postural sway was greater among individuals with joint hypermobility compared to those without joint hypermobility. The increased postural sway among those with joint hypermobility suggests that an increase in range of joint motion may decrease the ability to maintain a basic, static position of eyes open, double-legged stance. When input is eliminated from certain visual, vestibular, and/or somatosensory systems, sway is increased making it more difficult to maintain static balance for those with joint hypermobility. These studies support the assessment of postural sway and interventions to enhance neuromuscular function and balance among individuals with joint hypermobility. Future studies should identify consistent joint hypermobility definitions by age group, examine the association of joint hypermobility and balance among adults over the age of 30 years, and consider longitudinal designs to examine the impact of joint hypermobility on postural sway and balance over time.

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Table 1: Article Comparisons

Author (Year)	Number of Subjects	Hypermobility Status	Age (In Years)	Other Subject Characteristics	Time and Pertinent Conditions Tested	Results
Iatridou[9] (2014)	41 total	21 subjects with JHS BHJMI >4/9 20 subjects without JHS BHJMI <u><</u>4/9	JHS Avg. 21.7 Without JHS Avg. 21.5	All subjects were female All subjects were Caucasian Subjects were not involved in organized sports participation or intensive motor activities	20 second test time Single-legged stance on dominant leg with eyes open (EO) Single-legged stance on dominant leg with eyes closed (EC) Single-legged stance on dominant leg with eyes open with head extended (EO-HE)	**Mediolateral Sway:** Statistically significant greater sway for JHS in EO compared to control • $p < 0.01$ Statistically significant greater sway for JHS in EO-HE conditions compared to control • $p < 0.05$ Sway was greater for JHS in EC condition however, not statistically significant **Antero-posterior Sway:** Statistically significant greater sway for JHS in EO-HE conditions compared to control • $p < 0.001$ Sway was greater for JHS in EO and EC conditions however, not statistically significant **Overall Sway:** Not assessed
Mebes[10] (2008)	31 total	13 subjects with hypermobility BHJMI <u>></u>6/9 18 subjects with	Hypermobility Avg. 28.1 Normal mobility	All subjects were female All hypermobile subjects met	15 second test time Single-legged stance on right leg with eyes open	**Mediolateral Sway (mm):** Statistically significant greater sway for hypermobile group compared to control • Hypermobile group: mean 4.8 +/-

		normal mobility BHJMI <6/9	Avg. 27.2	criteria 4 & 5 of the BHJMI system for the right lower extremity		0.6 SD • Control group: 4.3 mean +/- 0.9 SD 4.3 • p = 0.034 **Antero-posterior Sway:** No statistically significant difference between sway for hypermobile group compared to control **Overall Sway:** Not assessed
Schmidt[11] (2017)	132 total	96 subjects without GJH BHJMI <4/9 36 subjects with GJH BHJMI ≥4/9 21 subjects with GJH BHJMI ≥5/9 9 subjects with GJH BHJMI ≥6/9	Without GJH 14.1 GJH 4/9 13.9 GJH 5/9 13.9 GJH ≥6/9 14.2	All subjects were elite level athletes participating in ballet, gymnastics or handball	30 second test time Double-legged stance with eyes open (2EO) Double-legged stance with eyes closed (2EC) Single-legged stance on non-dominant leg with eyes open (1EO) Single-legged stance on non-dominant leg with eyes closed (1EC)	**Mediolateral Sway (mm):** Statistically significant greater sway for GJH 5 with 2EO compared to control • GJH 5: 3.0 mean +/- 0.7 SD • No GJH 5: 2.7 mean +/- 0.7 SD • p = 0.043 Statistically significant greater sway for GJH 4 with 2EC compared to control • GJH 4: 3.9 mean +/- 0.8 SD • No GJH 4: 3.6 mean +/- 0.8 SD • p = 0.001 Statistically significant greater sway for GJH 5 with 2EC compared to control • GJH 5: 4.0 mean +/- 0.7 SD • No GJH 5: 3.67 mean +/- 0.8 SD • p = 0.01 Statistically significant greater sway for GJH 4 with 1EO compared to control • GJH 4: 3.8 mean +/- 0.4 SD • No GJH 4: 3.4 mean +/- 0.5 SD • p = 0.002

						Statistically significant greater sway for GJH 4 with 1EC compared to control • GJH 4: 5.1 mean +/- 0.6 SD • No GJH 4: 5.1 mean +/- 0.9 SD • p = 0.02 Statistically significant greater sway for GJH 5 with 1EC compared to control • GJH 5: 5.2 mean +/- 0.4 SD • No GJH 5: 5.1 mean +/- 0.9 SD • p = 0.02 **Antero-posterior Sway (mm):** Statistically significant sway for GJH 4 with 2EO compared to control • GJH 4: 3.2 mean +/- 1.0 SD • No GJH 4: 2.6 mean +/- 0.7 SD • p = 0.03 Statistically significant sway for GJH 6 with 2EO compared to control • GJH 6: 3.7 mean +/- 1.4 SD • No GJH 6: 2.7 mean +/- 0.8 SD • p = 0.03 Statistically significant greater sway for GJH 4 with 2EC compared to control • GJH 4: 4.0 mean +/- 0.7 SD • No GJH 4: 3.5 mean +/- 0.9 SD • p < 0.001 Statistically significant greater sway for GJH 5 with 2EC compared to control • GJH 5: 4.0 mean +/- 0.6 SD • No GJH 5: 3.6 mean +/- 1.0 SD • p = 0.001 No statistically significant greater sway

						for GJH with 1EO was found compared to control Statistically significant greater sway for GJH 5 with 1EC compared to control • GJH 5: 8.2 mean +/- 1.4 SD • No GJH 5: 7.7 mean +/- 2.0 SD • p = 0.047 **Overall Sway (mm):** Statistically significant greater sway for all test conditions GJH 4 compared to control 2EO: • GJH 4: 66.6 mean +/- 10.6 SD • No GJH 4: 57.2 mean +/- 11.6 SD • p = 0.001 2EC: • GJH 4: 98.1 mean +/- 19.1 SD • No GJH 4: 83.0 mean +/- 18.9 SD • p < 0.001 1EO: • GJH 4: 154.7 mean +/- 25.6 SD • No GJH 4: 142.1 mean +/- 29.8 SD • p < 0.001 1EC: • GJH 4: 288.3 mean +/- 49.9 SD • No GJH 4: 266.8 mean +/- 60.7 SD • p < 0.001

						Statistically significant greater sway for all test conditions GJH 5 compared to control 2EO: • GJH 5: 68.8 mean +/- 11.7 SD • No GJH 5: 58.1 mean +/- 11.4 SD • p = 0.001 2EC: • GJH 5: 104.3 mean +/- 17.4 SD • No GJH 5: 83.9 mean +/- 18.8 SD • p < 0.001 1EO: • GJH 5: 155.4 mean +/- 27.8 SD • No GJH 5: 143.7 mean +/- 29.1 SD • p < 0.001 1EC: • GJH 5: 281.9 mean +/- 34.1 SD • No GJH 5: 270.9 mean +/- 61.3 SD • p = 0.04 Statistically significant greater sway for 2EO and 2EC test conditions GJH 5 compared to control (exception 1EO condition) 2EO: • GJH 6: 70.5 mean +/- 12.9 SD • No GJH 6: 59.0 mean +/- 11.6 SD • p = 0.005 2EC:

						• GJH 6: 103.8 mean +/- 17.9 SD • No GJH 6: 85.9 mean +/- 19.7 SD • $p < 0.001$ 1EC: • GJH 6: 294.5 mean +/- 29.0 SD • No GJH 6: 270.8 mean +/- 59.9 SD • $p = 0.02$
Aydin[12] (2017)	69 total	29 subjects not hypermobile BHJMI ≤2/9 13 subjects moderately hypermobile BHJMI = 3-4/9 27 subjects distinctly hypermobile BHJMI ≥5/9	Not hypermobile Avg. 21.6 Moderately hypermobile Avg. 21.2 Distinctly hypermobile Avg. 20.8	All subjects were female	32 second test time Double-legged stance with neutral head on a firm surface with eyes open (NO) Double-legged stance with neutral head on a firm surface with eyes closed (NC) Neutral head on an elastic surface with eyes open (PO) Neutral head on an elastic surface with eyes closed (PC) Head turned 45 degrees right on a firm surface with eyes closed (HR) Head turned 45 degrees left on a firm surface with eyes closed (HL)	**Mediolateral Sway:** Not assessed **Antero-posterior Sway:** Not assessed **Overall Sway (mm):** Statistically significant sway for distinctly hypermobile group in HR compared to the non hypermobile group • 18.3 Mean +/- 6.0 SD • $p = 0.044$ Statistically significant sway for distinctly hypermobile group in HB compared to the non hypermobile group • Minimum 16.90 to Maximum 26.90 • $p = 0.038$

					Head extended about 30 degrees on a firm surface with eyes closed (HB) Head flexed about 30 degrees on a firm surface with eyes closed (HF)	
Juul-Kristensen[13] (2015)	27 total	16 subjects with GJH BHJMI ≥6/9 At least one hypermobile knee 11 subjects not GJH BHJMI ≤5/9 No hypermobile knee	GJH Avg. 14 Not GJH Avg. 14.3	All subjects were female	60 second test time Double-legged stance with eyes open (2EO) Double-legged stance with eyes closed (2EC) One-legged stance with eyes open on dominant leg (1EO)	**Mediolateral Sway:** Not assessed **Antero-posterior Sway:** Not assessed **Overall Sway (mm):** Statistically significant greater sway for GJH in 2EO compared to control group • GJH: 1.10 mean +/- 0.12 SD • No GJH: 0.99 mean +/- 0.13 SD • $p = 0.04$ Statistically significant greater sway for GJH in 2EC compared to control group • GJH: 1.64 mean +/- 0.29 SD • No GJH: 1.37 mean +/- 0.24 SD • $p < 0.001$

Legend:
JHS = Joint Hypermobility Syndrome, BHJMI = Beighton and Horan Joint Mobility Index, EO = Eyes Open, EC = Eyes Closed, EO-HE = Eyes Open with Head Extended, Avg. = Average, GJH = Generalized Joint Hypermobility, 2EO = Double-legged Stance with Eyes Open, 2EC = Double-legged Stance with Eyes Closed, 1EO = Single-legged Stance on Non-Dominant Leg with Eyes Open, 1EC = Single-legged Stance on Non-Dominant Leg with Eyes Closed, NO = Double-legged Stance with Neutral Head on a Firm Surface with Eyes Open, NC = Double-legged

Stance with Neutral Head on a Firm Surface with Eyes Closed, PO = Neutral Head on an Elastic Surface with Eyes Open, PC = Neutral Head on an Elastic Surface with Eyes Closed, HR = Head Turned 45 Degrees Right on a Firm Surface with Cyes closed, HL = Head Turned 45 Degrees Left on a Firm Surface with Eyes Closed, HB = Head Extended about 30 Degrees on a Firm Surface with Eyes Closed, HF = Head Flexed about 30 Degrees on a Firm Surface with Eyes Closed

Table 1: Beighton and Horan Joint Mobility Index (BHJMI)

a) Maneuver 1: Passively extend each little finger
One point given for each little finger extending beyond 90 degrees

b) Maneuver 2: Passively oppose each thumb to the forearm
One point given for each thumb contacting the forearm

c) Maneuver 3: Passively extend each elbow
One point given for each elbow in at least 10 degrees of hyperextension

d) Maneuver 4: Passively extend each knee
One point given for each knee in at least 10 degrees of hyperextension

e) Maneuver 5: Forward flex trunk to place the hands flat on the ground
One point given for the ability to place hands flat on the floor while knees are straight

Figure 2: Brighton Criteria[8]

- Major Criteria
  - BHJMI Score >4/9 (either currently or historically)
  - Arthralgia for longer than 3 months in 4 or more joints
- Minor criteria
  - A BHJMI score of 1,2, or 3/9 (0, 1, 2 or 3 if aged 50+)
  - Arthralgia ($\geq$3 months) in 1-3 joints, or back pain ($\geq$3 months), spondylosis, spondylolysis/spondylolisthesis
  - Dislocation/subluxation in more than one joint, or in one joint on more than one occasion
  - Soft tissue rheumatism $\geq$3 lesions (e.g. epicondylitis, tenosynovitis, bursitis)
  - Marfanoid habitus (tall, slim, span/height ratio > 1.03, upper:lower segment ration < 0.89, arachnodactyly (+ Steinberg/wrist signs)
  - Abnormal skin: striae, hyperextensibility, thin skin, papyraceous scarring
  - Eye signs: drooping eyelids or myopia antimongoloid slant
  - Varicose veins or hernia or uterine/rectal prolapse
- Requirement for Diagnosis
  - Any one of the following:
    - Two major criteria
    - One major plus two minor criteria
    - Four minor criteria
    - Two minor criteria and unequivocally affected first-degree relative in family history

Figure 3: Search Strategy