Mitral Valve Prolapse In Patients With Benign Joint Hypermobility Syndrome (BJHS)

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ABSTRACT

Background: Joint hypermobility was first mentioned by Hippocrates as an isolated feature, when he described the Celts' Incapacity to Pull a Bowstring or Throw a Dart, Due to The Slackness of Their Limbs

Objective: to determine the prevalence of mitral valve prolapse(MVP)in patients with benign hypermobility syndrome (BJHS).

Type of the study: Cross -sectional study.

Methods: Ninety patients with BJHS were included in this study. Full cardiological assessment was done for all of them, which include clinical examination, electrocardiography and echocardiography. Cardiac assessment was done for another sixty age and sex matched (normal mobile) Individuals served as a control group. Statistical analysis was done by using T test or chi square as indicated.

Results: Among 90 patients with BJHS, MVP was reported in 26 patients (28.9%) compared to four individuals (6.7%)of the control group on modern echocardiography studies (P=0.013).

Conclusions: the prevalence of MVP was significantly higher among patients with BJHS compared to normal mobile individuals.

Key Words: Mitral valve prolapse, Benign hypermobility syndrome

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Joint hypermobility (JH) was first mentioned by Hippocrates as an isolated feature, when he described the Celts' Incapacity to Pull a Bowstring or Throw a Dart, Due to The Slackness of Their Limbs (1). Hypermobility Syndrome Is Commonly Occurring and Frequently Overlooked Entity, Which Causes Much Suffering in The Community (2). JH Arouse Great Interest in The Last Decades for Being Associated to Musculoskeletal Dysfunctions, As Well As Abnormalities in Several Organic Systems and Among Them, The Mitral Valve Prolapse (MVP) (3).

Joint Hypermobility (JH) Is Defined As The Capacity To Perform Articular Movement With Higher Range Of Motion The Normal One(4)(5).Its Prevalence Depend On The Characteristics Of The Studied Group Determined ,Among Others ,By Age ,Sex, Ethnicity And Genetic Factors That Have An Effect On The Process Of Movement Evolutions .Studies Have Stated That Female Individuals Have A Higher Joint Mobility Than Male Individuals And That It Decreases With Age (6)(7).Approximately 30% Of The Adult Individuals Are Considered As Presenting JH(8). The Feature of JH That Appear in Some Hereditary Disease Such as Marfan Syndrome, Osteogegesis Imperfecta, Achard Syndrome, Homocystinuria And Hyperlysinemia Must Be Differentiated from Those Present in Normal Population.

JH in the general population is a common condition and a genetically determined one (8). There is a new consensus that the Benign Joint Hypermobility Syndrome (BJHS) is a multisystemic disorder, of which characteristics coincide with the characteristics of the hereditary disorders of the connective tissue, which include Marfan syndrome, Ehlers-Danlos syndrome and *Osteogenesis Imperfecta* (9).

The MVP is a common cardiovascular disorder, with a strong hereditary component, generally diagnosed in young individuals (10). Mitral valve prolapse was reported during clinical practice in patients with various heritable disorder of connective tissue such as Ehlers-danlose syndrome (12), Marfan's syndrome (13), and Osteogenesis imperfecta (14).

There were conflicting report regarding the association between BJHS and MVP. Some reports show increase incidence (14,15,16,17), and other challenged this association and show no difference between the incidence of MVP in BJHS and normal mobile subjects (18,19).

Methods: Ninety patients with joint hypermobility were included in this cross-sectional study.

All patients were evaluated for cardiac and respiratory symptoms and signs. Full cardiological evaluation was done blindly by another physician in all patients, which include clinical examination, electrocardiography and echocardiography, which was performed by using two-dimension voluson 5300 D-Kretz technik -S-PPA2-4 with a 2.5MHz probe.

During examination, particular attention was paid to the presence or absence of echocardiographic evidence of MVP according to the echocardiographic diagnostic criteria of MVP by Perloff et al (20), Cardiac assessment was done for another sixty age and sex matched (normal mobile) individuals served as a control group.

Statistical analysis, the T-test was used for mean age and chi-square for other variable correlation.

Results: All the 90 hypermobile patients (75 females and15 males), age range from 11 to 40 years with a mean of 22.68+_5.69 and 60 normal mobile individuals (50 females and 10 males). The demographic finding of patients with BJHS and controls are shown in table (1).

Among the hypermobile group, mitral valve prolapse was diagnosed in 26 patients (4 males and 22 females), two of them with mild mitral regurgitation,5 patients had murmurs on auscultation and another six with mid systolic click. Four individuals of the control group had

mitral valve prolapse (6.7%) on echocardiography. So, MVP was reported significantly higher among patients with BJHS compared to the control group (P=0.013).

Table (1): Demographic findings of 90 patients with BJHS and 60 controls.

Findings	Patients	Control
Mean age	22.68±56.9(11-40) year	22.17±4.98(11-40) year
Sex: Female	75	50
Male	15	10
Beighton score		
0-3	0	60
4	13	0
5-6	51	0
7-9	26	0
Mean Beighton score	5	1

Beighton score of ≥ 4/9 were reported among all the hypermobile group, and 54 patients (60%) had arthralgia for >3 months in more than four joints, 15 of them had MVP (57.7%), while 23 patients (25.6%) had arthralgia in 1-3 joints;9 of them with MVP (34.6%).

Marfanoid habitus was reported in 20 patients (22.2%), six of them was with MVP. Only 12 patients (13.3%) had past or present history of dislocation with three of them had MVP. Soft tissue rheumatism was reported in 8 patients (8.9%) and only 2 of them showed MVP.

Asthma and wheezy chest was reported only in 4 patients (4.4%), one of them only had MVP as shown in table (2).

Table (2): the correlation between various clinical findings and MVP in 90 patients with BJHS.

Clinical feature	No. of patients	%	No. of patients with MVP	PERCENT of clinical feature in patient with MVP
Joint mobility score ≥ 4	90	100	26	100
Arthralgia ≥4joints	54	60	15	57.7
Arthralgia 1-3joints	23	25.6	9	34.6
History of dislocation	12	13.3	3	11.5
Soft tissue rheumatism	8	8.9	2	7.7
Marfanoid habitus	19	21.1	6	23.1
Asthma & wheezy chest	4	4.4	1	3.8

Discussion: In this study, MVP was reported in 28.9% of hypermobile patients and 6.8% of controls (P=0.013). We have concentrated on the incidence of MVP. There were conflicting reports on the association between BJHS and MVP in various studies from different ethnic groups. MVP was considered in the 1980s to be feature of hypermobile, but this was questioned in the 1990s on using modern echocardiography technology and criteria. Graham, 1981(14), Pitcher,1982(15), Handler,1985(21), and Ondrasik,1988(22), all described an increase prevalence of MVP in individuals with joint hypermobility or vice versa. Other studies by Jessee,1980(18) and Marks, 1983(23) challenged this association and conclude in their studies that those patients with MVP do not have generalized joint laxity.

Mishra et al,1996(19) who used modern echocardiographic technique and strict criteria showed no evidence of increase of MVP among 58 hypermobile subjects compared with 30 normal mobile age and sex matched control.

There was a report from Venezuela by Riana et al,2001(24) and although they used the Beighton 9 score criteria and not the new set of criteria for BJHS. There was no mention of the type of echocardiogram used in the study, yet they have concluded that among extra-articular manifestations, MVP was commonest one.

Conclusion: The prevalence of MVP was significantly higher among our patients with BJHS compared to normal mobile individuals.

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