



## A CASE REPORT OF BENIGN HYPERMOBILITY SYNDROME IN A 42 YEAR OLD PATIENT

Gaurav Solanki\*<sup>1</sup>, Namita Lohra<sup>1</sup>, Jaya Lohra<sup>1</sup>, Renu Solanki<sup>2</sup>

<sup>1</sup>Jodhpur National University, Jodhpur, Rajasthan, India.

<sup>2</sup>Lachoo Memorial College of Science and Technology, Rajasthan, India.

Corresponding Author:- **Gaurav Solanki**  
E-mail: [drgauravsolanki@yahoo.com](mailto:drgauravsolanki@yahoo.com)

<p><b>Article Info</b> Received 22/10/2014 Revised 07/11/2014 Accepted 12/11/2014</p> <p><b>Key words:</b> Hypermobility, Syndrome, Rheumatic etc.</p>	<p><b>ABSTRACT</b> A case of Benign Hypermobility Syndrome (BHS) is discussed. The case was a young male aged 42 with musculoskeletal symptoms since 2 years. The patients had hypermobility of joints in absence of demonstrable systemic rheumatic disease. The prevalence, clinical features and management of the entity is discussed. An increased awareness of this condition among doctors is warranted as some patients may be erroneously diagnosed as RA/SLE and may be put on DMARDs and steroids.</p>
--	---

### INTRODUCTION

Generalized hyper mobility in the absence of systemic disease is a common condition that has a prevalence of 4 - 13% in the general population. Children have higher laxity of joints which diminishes gradually during adolescence and adulthood. At any given age, females have a greater degree of joint laxity. It also varies among ethnic groups – more common in Africans, Asians and Middle-East descent [1]. Hypermobility of joints is also seen in Marfan's Syndrome and Ehler Danlos Syndrome (EDS) and hence the prefix benign to distinguish from the latter. There are many common features between EDS and Benign Hyper mobility Syndrome (BHS). The condition had been first described by Hippocrates in Scythian warriors, who were warrior tribesmen from Central Asia who invaded India and established their kingdoms. Detailed history, careful clinical examination and the modified Brighton score aid in the accurate diagnosis [2].

### CASE REPORT

A 42 year old male presented with multiple joint pains and low backache since 1 year. The pain occurred at any time of the day and even occurred at night. There were

2 - 4 attacks of severe pain in a month, lasting for 1 - 2 hours, responding to analgesics and rest. Pain did not occur daily and some days were better than the others. Backache was not aggravated by bending forwards, coughing, sneezing or straining. He had been diagnosed as Spondyloarthropathy and had been on various NSAID's and DMARD's (hydroxychloroquine, sulphasalazine), steroids and calcium supplements, with no relief. On examination of the musculoskeletal system, there was no joint swelling, tenderness, deformity or restriction of movement; no tender points for fibromyalgia were 16 out of 18. Complete back flexion was possible; Schoeber's test was negative. There was no tenderness of sacroiliac joints. There was hypermobility of many joints and the Beighton Score was 8/9. Haemogram and biochemical tests were within normal limits. All radiographs of various joints, X-Ray and MRI of sacroiliac joint were normal. On the basis of clinical findings, Beighton score of 8/9 and normal investigations, a diagnosis of BHS was made. The DMARD's were omitted, he was counseled, advised supervised physiotherapy and he is doing well.



## DISCUSSION

Diagnosis of BHS is one of the very difficult tasks. When patients present with pain in multiple joints with hypermobility in multiple joints; systemic rheumatic diseases should first be excluded. Some patients also have localized hypermobility, which can make the diagnosis difficult. There is also considerable variation between different ethnic groups. Modified Beighton score is widely accepted as a measure of joint hypermobility and assesses nine genetically determined sites. The testing involves 5 simple bedside clinical maneuvers and giving a score. A Beighton score of 4 or more is indicative of general hypermobility. Revised Brighton criteria, Bulbena criteria are some others. Careful history and clinical examination is essential as many patients with hypermobility have positive rheumatoid factor or antinuclear antibodies and are misdiagnosed as rheumatoid arthritis or SLE [3,4].

The pain associated with it is related to recurrent unrecognizable trauma. The ligament laxity and joint instability leads to recurrent dislocations, subluxations and sprains. Some develop correctable deformities, including swan – neck deformities of fingers, hallux valgus and varus deformities of feet in absence of arthritis, but can be mistaken as rheumatoid arthritis. Because joints stiffen with age, the natural history of BHS is typically one of improvement, with progressively lessening degrees of joint laxity and associated musculoskeletal symptoms. Many affected children outgrow their symptoms during adolescence or adulthood, and women may experience fewer symptoms following menopause. However, over many years, joint hypermobility can predispose to osteoarthritis, even prematurely, in the 4<sup>th</sup> or 5<sup>th</sup> decade followed by eventual chondrocalcinosis. BHS can also be associated with fibro-myalgia syndrome [5, 6].

For acute symptoms of this disease, NSAID's may be used [7]. Regarding chronic management, patients should be advised to identify the activities that precipitate symptoms and modify their lifestyles accordingly. While on one hand, the precipitating and repetitive activities such as certain sports or hobbies should be avoided, by no means inactivity should be encouraged. Moderate exercise is

extremely beneficial, by maximizing the muscle support around the hyper mobile joint. Exercise therapy should aim at improving muscle tone, joint stability and proprioception. But it should be under the supervision of a physiotherapist familiar with the disorder, tailored to suit the patient's needs, as inappropriate use can exacerbate the symptoms. Isometric exercises should be advised that improve muscle tone without increasing the joint instability. Because the knee joint is commonly involved, quadriceps exercises can be especially helpful. Closed kinetic chain exercises improve proprioception. In these exercises, the exercising limb, visualized as a chain of jointed segments, is the kinetic chain. In closed kinetic chain, the ends of the chain are fixed, while the intervening segments make small controlled movements [8,9]. Improved muscle tone and coordination and joint proprioception helps in avoiding injuries. Patients with back pain benefit from back strengthening exercises and improved posture. Most patients need sympathetic counseling and explanation. Reassurance that they are not suffering from serious diseases like RA and SLE will alleviate considerable suffering. Nearly 5% of normal individuals are positive for R factor; it often leads to misdiagnosis and unnecessary treatment. Awareness of high prevalence of the condition, a good history, clinical examination needs to be over and re-emphasized. Nevertheless, some patients may develop inflammatory disease later, so regular follow up is required [10].

## CONCLUSION

The hands with hyper extensile joints may mimic deformities of Rheumatoid Arthritis, but they are completely reducible deformities and there is no stiffness, swelling or tenderness. As Doctors, we need to understand the impact that BHS may have on many patients for whom each day is a struggle against pain and injury. Reassure the patients we must, but also accept the fact that we, the medical community have not really been able to provide effective cure for them.

## REFERENCES

1. Zweers MC, Dean WB, van Kuppevelt TH, Bristo WJ, Schalkwijk J. (2005). Elastic fiber abnormalities in hypermobility-type of Ehlers-Danlos syndrome patients with tenascin-X mutations. *Clin Genet*, 67, 330 - 4.
2. Ferrell WR, Tennant N, Sturrock RD, Ashton L, Creed G, Brydson G, Rafferty D. (2004). Amelioration of symptoms by enhancement of proprioception in patients with joint hypermobility syndrome. *Rthritis rheum*, 50, 33 - 8.
3. Lawrence A. (2005). Benign Hypermobility Syndrome. *J Rheumrol Assoc*, 13, 150 - 5.
4. Beighton P, Solomon L. (1973). Articular mobility in an African population. *Ann Rheum Dis*, 32, 413 - 8.
5. Bulbena A, Duro JC, Porta M, Faus S, Vallescar R, Martin R. (1992). Clinical assessment of hypermobility of joints, assembling criteria. *J Rheumatol*, 19, 115 - 22.
6. Everman DB. (1998). Hypermobility Syndrome, A review. *Paediatrics in Review*, 20, 9 - 17.
7. Borghetti A, Gardella J. (1990). Hypermobility syndrome. *The International journal of periodontics & restorative dentistry*, 10(3), 216.
8. Jahnke P, Sandifer J, Gher M, Gray J, Richardson A. (1993). Hypermobility. *Journal of periodontology*, 64(4), 315.
9. Everman DB, Robin NH. (1998). Hypermobility Syndrome. *Paediatrics in Review*, 19, 111 - 7.
10. Grahame R, Edwards JC, Pitcher D, Gabell A, Harvey W. (1981). A clinical and echocardiographic study of patients with the hypermobility syndrome. *Ann Rhem Dis*, 40, 541 - 6.

