

# Ethnicity, hypermobility syndrome and Ehlers-Danlos syndrome: A study based on hospital admissions

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## ABSTRACT

The purpose of the study was to identify any evidence of different patterns of disease for Ehlers Danlos Syndrome (EDS) and Hypermobility Syndrome (JHS) in the White British and South Asian communities in England. The numbers of hospital in-patients in ten NHS Trusts, between 2016 and 2020, in whom a diagnosis of EDS or JHS had been recorded were identified through Freedom of Information (FOI) requests. The condition was either the primary diagnosis or a secondary one, incidental to the reason for admission. The ten Trusts were selected because they served areas with a significant South Asian population and had been involved in previous studies of discrimination in the delivery of care. The study also assessed whether there was evidence of any of the Trusts underserving their South Asian community with lesser levels of recognition of EDS and JHS. Overall South Asian patients were almost four times more likely to have been diagnosed as having JHS. This proportion was statistically significant ( $z = -11.69$ ,  $p < 0.00001$ ). In Burton and Derby, Cambridge and Leicester the proportion of South Asian patients diagnosed as having EDS was significantly lower than in other Trusts ( $z = 9.4$ ,  $p < 0.00001$ ). This was also the case for JHS ( $z = 8.09$ ,  $p < 0.00001$ ). This would indicate that in these three Trusts both conditions are underdiagnosed in the South Asian community. The significance of these findings is discussed.

**Keywords:** Ehlers-Danlos syndrome, hypermobility syndrome, South Asian ethnicity

## INTRODUCTION

During the last decade Ehlers-Danlos Syndrome (EDS) has been recognized more frequently by clinicians. During this time the nature and classification of Joint Hypermobility Syndrome (JHS) has changed. After 2017, the term JHS was dropped and patients are described as having either hypermobility spectrum disorder (HSD) or hypermobile EDS (hEDS) based on international criteria [1]. However, in practice many hospital units still classify patients according to the older definitions.

As to the magnitude of the problem, two large population studies have looked at the prevalence of JHS and EDS. One specifically looked at the prevalence of these conditions [2]; the second looked at mental health and neurodiversity. However, these data can be used to estimate the prevalence of JHS and EDS [3]. A third study also gives a sense of the

prevalence of JHS by extrapolating observations from rheumatology clinics in the UK [4]. Overall, these studies suggest that the combined prevalence of JHS and EDS is between 125 and 200/105 population. The prevalence of JHS probably lies between 111 and 167/105 population, whilst the range for EDS is between 20 and 33/105 population [5]. It is likely that these figures significantly underestimate the true prevalence of EDS, as many people are either not diagnosed or misdiagnosed.

Against this background, the Ehler-Danlos Society in the United Kingdom sought to promote better understanding of this disease in the South Asian community by translating its literature into a range of appropriate languages. However, apart from sporadic case reports, little has been published on this condition in either the Indian sub-continent or the South Asian diaspora. This contrasts strikingly with inflammatory bowel disease [6]. Similarly, although there is

widespread recognition of the stigmata associated with mental welfare problems in the South Asia community [7], limited attention has been given to chronic diseases with an inherited element. It is likely that the severity of chronic conditions will play a significant part in the readiness of the South Asian community to accept the diagnosis and directly affect someone's marital prospects and, indeed, that of other family members [8].

Against this background the purpose of this study was to gain some idea of the magnitude of the condition in the South Asian community in England and to determine whether JHS was a preferred diagnosis. The study was not intended to be an epidemiological assessment but rather to seek evidence of whether there were differences between the South Asian and White British communities. No attempt was undertaken to identify causes for any such differences.

## METHOD

Ten NHS Trusts which served significant South Asian populations were identified. Each Trust was sent a Freedom of Information (FOI) request to provide data on patients who were hospital inpatients and also had a diagnosis of Ehlers Danlos Syndrome (Q79.6) or Hypermobility syndrome (M35.7) between 2016 and 2020. This, therefore, included patients who were admitted for reasons other than arising directly from these two conditions. Data were to be provided separately for patients who were:

1. White British (National Code A)
2. Asian (Pakistani, Indian and Bangladeshi origin - National Codes H, J and K).

The proportion of these patients, who had EDS, was compared between communities, using a z statistic [9].

## RESULTS

The ten Trusts provided the requested data (See Table). Comparable numbers of White British patients were admitted with EDS or JHS. However, overall South Asian patients were almost four times more likely to have been diagnosed as having JHS than EDS. This proportion was statistically significant ( $z = -11.69$ ,  $p < 0.00001$ ).

Within individual Trusts, this difference in the proportion of patients was not always seen. For example, in Trusts where small number of patients

**TABLE 1.** Inpatients with a diagnosis of Ehlers Danlos Syndrome (EDS) or Hypermobility Syndrome in Ten NHS Trusts in England between 2016 and 2020

NHS Trust	EDS		Hypermobility	
	White British	South Asian	White British	South Asian
Derby & Burton	517	11	766	6
<b>Birmingham</b>	221	<15	206	<19
Bradford	223	12	435	130
Cambridge	1424	6	584	1
Croydon	71	1	43	8
<b>East Lancashire</b>	223	39	486	155
Barts	247	25	162	49
Redbridge	60	1	116	10
Wolverhampton	216	0	533	19
Leicester	187	8	162	4
Total	3389	117	3493	400

were admitted, who were noted to have either EDS or JHS as a primary or secondary diagnosis. this was not the case. In Burton and Derby, Cambridge and Leicester more patients had a diagnosis of EDS than JHS. However, in these trusts the proportion of South Asian patients in the combined figure for Codes A, H, J and K, diagnosed as having EDS was significantly lower than in the other Trusts ( $z = 9.4$ ,  $p < 0.00001$ ). This was also the case for JHS ( $z = 8.09$ ,  $p < 0.00001$ ). This would suggest that in these three Trusts both conditions are underdiagnosed in the South Asian community.

## DISCUSSION

EDS is seen in the South Asian community, but less likely to be diagnosed than JHS. Indeed, in some Trusts both diagnoses seem to be made at a much lower frequency than in others. The reasons for these differences could include:

1. A lower incidence of both diseases in the South Asian community, with different spectra for the diseases.
2. A reluctance amongst potential patients from the South Asian community to be diagnosed with a chronic incurable disease.
3. Less readiness amongst clinicians to make these diagnoses in South Asian patients. This could reflect a lack of awareness that the disease occurs amongst South Asians or discrimination in the delivery of health care. The findings from Leicester, Cambridge and Derby would suggest that discrimination does play a role in some Trusts and is supported by similar

issues in relation to inflammatory bowel disease [10,11].

This study was conducted using data obtained through FOI requests. This approach has been used in a number of earlier studies and the data have been shown to be robust [10,11]. There are, of course, weaknesses in such an approach and these have been well documented [12-14]. Perhaps the most important limitations are the enthusiasm with which the FOI officer approaches such requests and the accuracy of the data recorded by coding clerks at the relevant hospitals [15].

As yet, reports on EDS and JHS in the South Asian community in the UK and the Indian sub-continent

have been restricted to case reports. There is a clear need to obtain robust data on the incidence of these diseases in both the White British and South Asian communities. However, at present such work is limited by the changing definitions around these conditions. There is a clear need to educate clinicians better about their occurrence in both communities. In addition, the perceived stigma attached to such diagnoses needs to be addressed, especially in the South Asian community. However, it is likely that overall the underlying reasons will be little different to that for other chronic conditions, such as epilepsy and inflammatory bowel disease and, as yet, no effective solutions to such problems have been identified.

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## REFERENCES

1. Malfait F, Francomano C, Byers P, Belmont J, Berglund B, Black J, et al. The 2017 international classification of the Ehlers-Danlos syndromes. *Am J Med Genet C Semin Med Genet.* 2017 Mar;175(1):8-26. doi: 10.1002/ajmg.c.31552. PMID: 28306229.
2. Demmler JC, Atkinson MD, Reinhold EJ, Choy E, Lyons RA, Brophy ST. Diagnosed prevalence of Ehlers-Danlos syndrome and hypermobility spectrum disorder in Wales, UK: a national electronic cohort study and case-control comparison. *BMJ Open.* 2019 Nov 4;9(11):e031365. doi: 10.1136/bmjopen-2019-031365. PMID: 31685485; PMCID: PMC6858200.
3. Cederlöf M, Larsson H, Lichtenstein P, Almqvist C, Serlachius E, Ludvigsson JF. Nationwide population-based cohort study of psychiatric disorders in individuals with Ehlers-Danlos syndrome or hypermobility syndrome and their siblings. *BMC Psychiatry.* 2016 Jul 4;16:207. doi: 10.1186/s12888-016-0922-6. PMID: 27377649. PMCID: PMC4932739.
4. Grahame R, Hakim AJ. Joint hypermobility syndrome is highly prevalent in general rheumatology clinics, presentation being gender, age and race-related. *Ann Rheum Dis.* 2006;65(Suppl 2):263.
5. Kulas Sjøborg ML, Leganger J, Quitzau Mortensen L, Rosenberg J, Burcharth J. Establishment and baseline characteristics of a nationwide Danish cohort of patients with Ehlers-Danlos syndrome. *Rheumatology (Oxford).* 2017 May 1;56(5):763-767. doi: 10.1093/rheumatology/kew478. PMID: 28077691.
6. Farrukh A, Mayberry JF. Inflammatory bowel disease and the South Asian diaspora. *JGH Open.* 2019 Feb 22;3(5):358-360. doi: 10.1002/jgh3.12149. PMID: 31633038. PMCID: PMC6788368.
7. Karasz A, Gany F, Escobar J, Flores C, Prasad L, Inman A, et al. Mental Health and Stress Among South Asians. *J Immigr Minor Health.* 2019 Aug;21(Suppl 1):7-14. PMID: 27848078. PMCID: PMC5643212. doi: 10.1007/s10903-016-0501-4.
8. Mayberry JF, Farrukh A. Gastroenterology and the provision of care to Panjabi patients in the UK. *Frontline Gastroenterol.* 2012 Jul;3(3):191-198. doi: 10.1136/flgastro-2012-100119. Epub 2012 Apr 13. PMID: 28839663. PMCID: PMC5517281.
9. Social Science Statistics. <https://www.socscistatistics.com/> (Accessed 15 October 2022)
10. Farrukh A, Mayberry JF. Ethnic variations in the provision of biologic therapy for Crohn's disease: a Freedom of Information study. *Med Leg J.* 2015 Jun;83(2):104-8. Epub 2015 Jan 7. PMID: 25568172. doi: 10.1177/0025817214563907.
11. Farrukh A, Mayberry JF. Evidence of on-going disparate levels of care for South Asian patients with inflammatory bowel disease in the United Kingdom during the quinquennium 2015–2019. *Gastrointestinal Disorders.* 2022 Jan 14;4(1):8-14. doi: 10.3390/gidisord4010002.
12. Swallow D, Bourke G. The freedom of information act and higher education: The experience of FOI officers in the UK. The Constitution Unit London, 2012. <https://www.ucl.ac.uk/constitution-unit/sites/constitution-unit/files/he-foiofficers-survey-report.pdf> (Accessed 22 October 2022).
13. Fowler AJ, Agha RA, Camm CF, Littlejohns P. The UK Freedom of Information Act (2000) in healthcare research: a systematic review. *BMJ Open.* 2013 Nov 27;3(11):e002967. PMID: 24285626; PMCID: PMC3845048 doi: 10.1136/bmjopen-2013-002967.
14. Bourke G, Worthy B, Hazell R (2012). Making freedom of information requests. A guide for academic researchers. London: The Constitution Unit, University College.
15. Burns EM, Rigby E, Mamidanna R, Bottle A, Aylin P, Ziprin P, et al. Systematic review of discharge coding accuracy. *J Public Health (Oxf).* 2012 Mar;34(1):138-48. Epub 2011 Jul 27. PMID: 21795302; PMCID: PMC3285117. doi: 10.1093/pubmed/fdr054.