Criterion 1: Generalized joint hypermobility

 Beighton score:

 > 6 in prepubertal children and adolescents

 > 5 in pubertal men and women up to age 50

 > 4 men and women over the age of 50

 If one point below cut off two or more "yes” answers to the five point questionnaire should be considered (Table 3).

 Criterion 2: Two or more of the following features (A, B, or C) must be present

 Feature A (live must be piescm):

 • Unusually soft or velvety skin

 • Mild skin hyperextensibility

 • Unexplained striae distensae or rubae at the back, groin, thighs, breasts and/or abdomen in adolescents, men or prepubertal women without a history of significant changes in weight

 • Bilateral piezogenic papules of the heel

 • Recurrent or multiple abdominal hernias

 • Atrophic scarring involving at least two sites without the formation of papyraceous and/or hemosideric scars

 • Pelvic floor, rectal and/or uterine prolapse in children, men or nulliparous women without history of morbid obesity or predisposing medical condition

 • Dental crowing and high or narrow palate

 • Arachnodactyly (positive Walker sign or Steinberg sign on both sides)

 • Arm span to height ratio >1.05

 • Mitral valve prolapse

 • Aortic root dilatation with Z-score >+2

 Feature B:

 • Positive family history (one or more first degree relatives meeting current criteria for hEDS)

 Feature C (must have at least one):

 • Musculoskeletal pain in two or more limbs, recurring daily for at least 3 months

 • Chronic, widespread pain for >3 months

 • Recurrent joint dislocations or joint instability in the absence of trauma

 Criterion 3: All of the following must be met

 • Absence of unusual skin fragility, which should prompt consideration of other types of EDS

 • Exclusion of other heritable and acquired connective tissue disorders. In patients with an acquired connective tissue disorder, additional diagnosis ofhEDS requires meeting both features A and B of

 criterion 3. Feature of criterion C cannot be counted in this situation

• Exclusion of alternative diagnoses that may also include joint hypermobility by means of hypotonia and/or connective tissue laxity. Alternative diagnoses may include: neuromuscular disorders (e.g:Bethlem myopathy), other hereditary connective tissue disorders (e.g., Loeys-Dietz syndrome, Marfan syndrome, other types of EDS) and skeletal dysplasias (e.g., osteogenesis imperfecta)