**Joint Hypermobility**

**Referral Guideline**

MAINE MEDICAL PARTNERS - PEDIATRIC SPECIALTY CARE (DIV. OF GENETICS) • 1577 CONGRESS ST, PORTLAND, ME • (207) 662-5522

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### High Risk

**Symptoms and Labs**
- Joint hypermobility in the presence of:
  - Personal history of aortic or other arterial aneurysm/rupture
  - Personal history of lens dislocation
  - Current pregnancy and there are additional symptoms as noted for moderate risk category (yellow box)

**Isolated joint hypermobility is not an indication for an emergent Genetics consultation**

**Suggested Previsit Workup**
- If currently pregnant obtain baseline or follow up echocardiogram

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### Moderate Risk

**Symptoms and Labs**
- Joint hypermobility in presence of:
  - Recurrent dislocations
  - Marfanoid body habitus
  - Excessively soft (doughy), stretchy skin
  - Skin fragility, tears
  - POTS (postural orthostatic tachycardia)
  - Excessive bruising in absence of known trauma
  - Documented family history of a known connective tissue disorder

**Suggested Workup**
- Referral encouraged

If there is known family history of a specific disorder, we will require documentation of DNA testing or how a specific connective tissue disorder diagnosis was established in an affected family member

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### Low Risk

**Symptoms and Labs**
- Asymptomatic joint hypermobility in the absence of skin or cardiovascular concerns, especially in young children, is low risk

**Suggested Management**
- Monitor
- Joint precautions as needed (e.g. If joint pain or injury is a concern)

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### Clinical Pearls

- **ALL referrals are reviewed by an ABGC Certified Genetic Counselor and/or a Geneticist; we will expedite any referral when medically indicated.**
- *Joint hypermobility is clinically defined as a Beighton score of > 5. The Beighton maneuvers are a validated examination screening procedure based on passive movements with one point scored for each of the following: Passive dorsiflexion of each 5th finger at MCP joint > 90°, Passive apposition of each thumb to flexor surface of forearm, Hyperextension of each elbow > 90°, Hyperextension of each knee > 90°, Ability to place palms on the floor with knees fully extended. Beighton et al. Ann Rheum Dis 1973;32:413-418.**
- Joint hypermobility is a common multifactorial trait influenced by age, conditioning, genetic background and gender.
- Joint pain may accompany joint hypermobility and is not necessarily an indicator of underlying connective tissue disorder.

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These clinical practice guidelines describe generally recommended evidence-based interventions for the evaluation, diagnosis and treatment of specific diseases or conditions. The guidelines are: (i) not considered to be entirely inclusive or exclusive of all methods of reasonable care that can obtain or produce the same results, and are not a statement of the standard of medical care; (ii) based on information available at the time and may not reflect the most current evidenced-based literature available at subsequent times; and (iii) not intended to substitute for the independent professional judgment of the responsible clinician(s). No set of guidelines can address the individual variation among patients or their unique needs, nor the combination of resources available to a particular community, provider or healthcare professional. Deviations from clinical practice guidelines thus may be appropriate based upon the specific patient circumstances.