

Hypermobility, Hyperlaxity, and Entesis

There are children and teenagers who have exaggerated but benign pains that are similar to growing pains. These pains which are often described as severe, can be present at any time of the day, and are typically increased by physical activity. The conundrum for parents is that the use of the words “pain” or “hurts” does not always imply a worrisome condition. The concept of “fatigue” is not in many children’s vocabulary. These children often have distinct physical exam findings that aid in diagnosis of this syndrome: hyper mobile joints, tenderness around tendons

and ligaments (Figure 1), lack of muscle soreness, absence of swelling inside the joint, or lack of atrophy of the muscles.

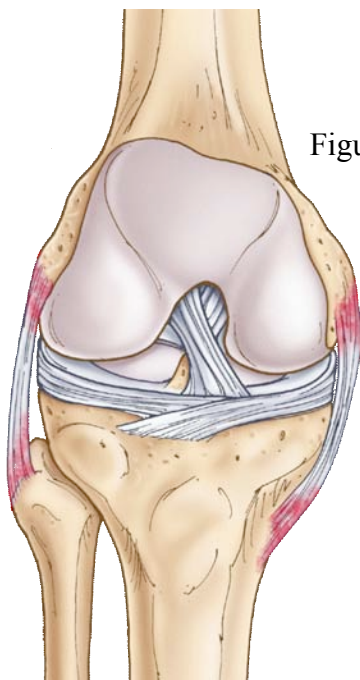


Figure 1



Figure 2

Hypermobility, either generalized or localized, is a common finding in children with pain. The diagnosis of hypermobility is considered if the patient can perform some of the following tests: push the thumb until it touches the forearm (Figure 2), hyperextension of the elbows (Figure 3) or pushing the knees backwards by more than 10 degrees (Figure 4), excessive dorsiflexion of the ankles or leaning forward and placing the hands flat on the floor.

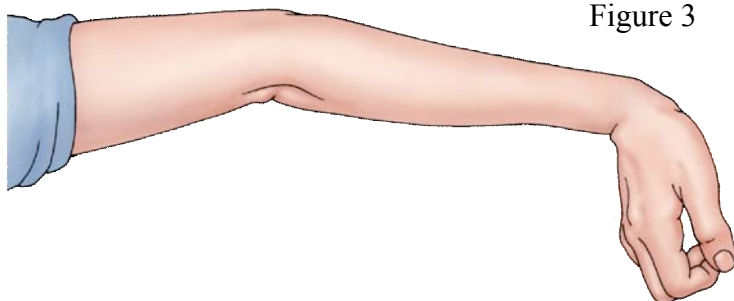
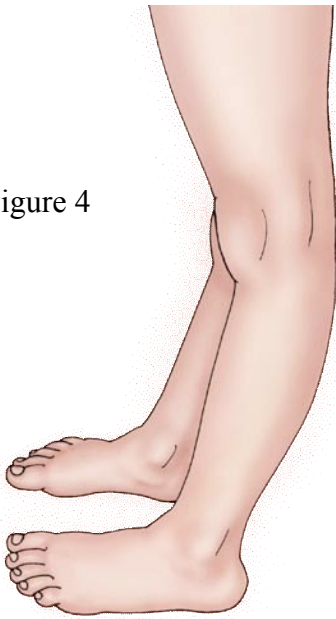


Figure 3

Figure 4



Severe hypermobility or hyperlaxity is a type of congenital disorder. In such conditions as Ehlers Danlos or Marfan's syndrome, the connective tissue, which is the basic building block for ligaments, joint capsule and tendons, is abnormal. Tendons and ligaments are made with a type of collagen that normally does not stretch or elongate. In these conditions the tissues stretch when walking or exercising abnormally. The result is that the muscles have to work "harder" and fatigue faster. In normal tendons the collagen fibers are like 2 by 4's arranged in such a manner as to make a steel cable for the muscle to move the bones. Between the muscle cells there are spaces. When a muscle contracts, the spaces are closed down, the muscle shortens and movement occurs.

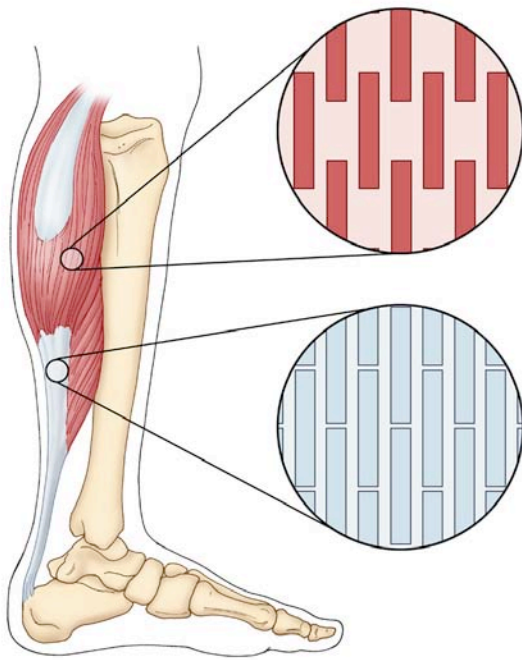


Figure 5: The muscle cells are separated from each at rest and the tendon cells are packed tightly together.

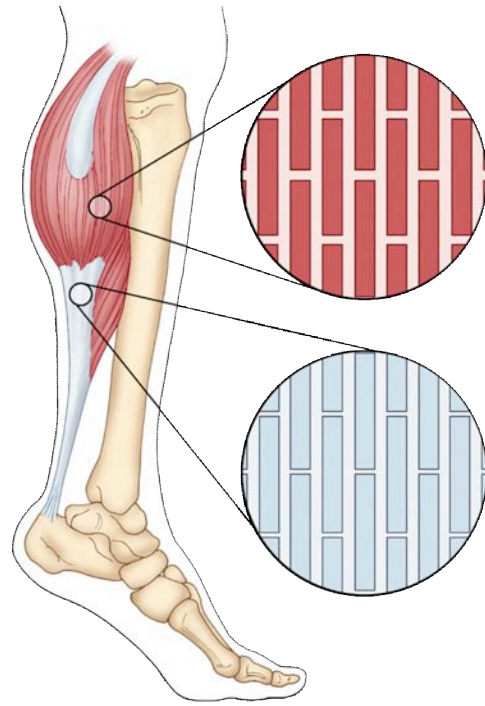


Figure 6: With muscle contraction, the muscle cells draw closer together. This shortening of the muscle results in the tendon being pulled and the bone moving.

With hypermobility, however, contraction of the muscle may not result in immediate movement because the tendons are not solid steel cables and some microscopic elongation occurs with movement. The muscle, therefore, may need to work harder in order to produce the same movement.

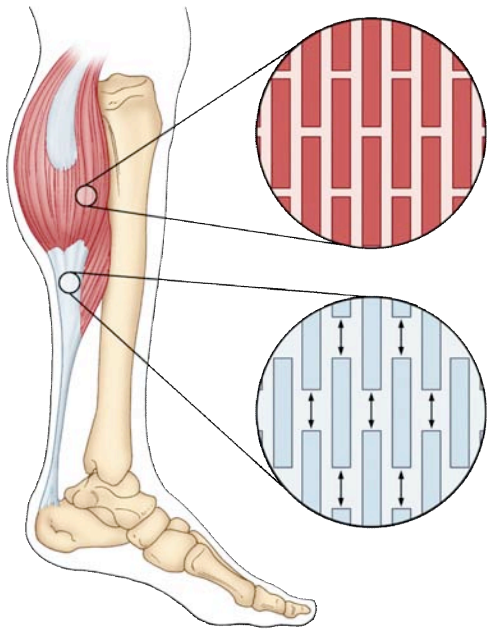


Figure 7: In Hypermobility, the tendon cells are not bound together tightly. The result is elongation of the tendon with muscle activity and increased energy expenditure.

The microscopic elongation of the tendons that occurs with activities has been described in a variety of ways. One example is the fatigue that is experienced when running in soft sand at the beach. Another example is that of train cars connected with rubber bands. The train cars do not move until all of

the stretch in the rubber band has occurred, but again it takes a lot more effort to move the cars because of the abnormal stretching of the links between the cars.

The other hallmark of hypermobility and hyperlaxity is the common complaint of extreme pain in the tendons and ligaments of the lower extremities. Patients with hypermobility do not typically feel the same “fatigue” or “muscle soreness” that normal people experience. The lack of stamina and the sensation of the legs buckling or collapsing when fatigued is all part of the clinical spectrum (Figure 8).

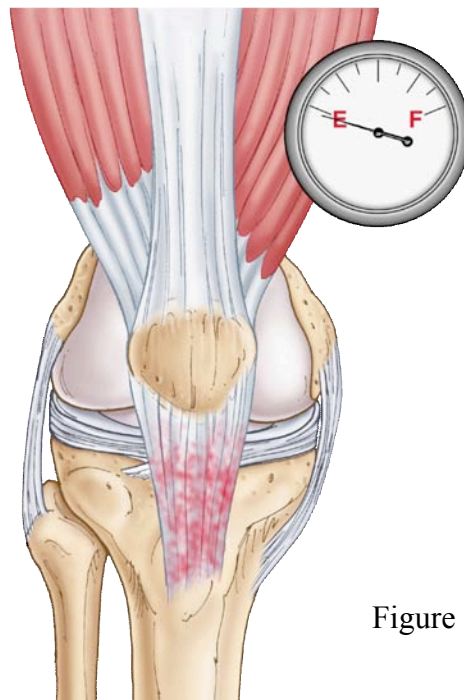


Figure 8

Occasionally laboratory and radiology studies are performed as part of the evaluation. In other cases, a medical genetics consultation is required when other diagnoses are more likely.

Activity modification, rest, nutrition and NSAID's are all important in minimizing the fatiguing effects of hypermobility. Year round sports, activities that involve prolonged training or running

seem to cause greater fatigue and perceived "pain" in children with hyperlaxity. The completion of the adolescent growth spurt corresponds to the production of adult hormones. While hormones cannot replace the defective tendons, they will produce muscle hypertrophy. The increase in size of the muscles usually leads to improved endurance.

