INTRODUCTION

Perthes disease is a disorder of the hip in young children in which the bone within the femoral head goes through a process of damage, collapse, and healing. It is not known why this process occurs. The process of damage occurs over several months, which is followed by collapse which occurs over several months, which is followed by healing which can take several years. This process, especially the healing part, always occurs. There are no easy answers for treatment. Younger children, age 6 and less, do very well in the long run. They are generally treated symptomatically with pain medicine and activity limitations. Occasionally, they need physical therapy, bracing, or casting. Older children, especially age 9 and old, can have lasting flattening of the femoral head which can lead to premature arthritis as an adult. Surgery is often recommended for these older children to minimize the risk for arthritis. Perthes Disease is also known by other names which include Legg-Calve-Perthes, Legg-Perthes, osteochondrosis of the hip, coxa plana, and idiopathic juvenile avascular necrosis of the hip.

CAUSES

The cause of Perthes disease is not known. There are scientists who believe the problem is related to hormonal imbalance, subtle coagulation differences, nutritional deficiencies, vascular occlusions, or repetitive mechanical stresses. It is likely that there is a combination of these factors which play different rolls in different patients. Many patients are male, thin and small for their age, and borderline hyperactive. The most common age is between 4 and 8 years of age, but it can occur as young as 2 and as old as 12. Various studies have reported that children with Perthes disease have a positive family history in 2 - 10% of the cases. Perthes disease is uncommon and affects about 1 in 20,000 people.

Some of the technical terms used to describe this condition include osteochondrosis and avascular necrosis. Osteochondrosis is the general term used to indicate problems with growing joint cartilage, such as in the elbow (Panner Disease), the knee (Blounts Disease and Osgood-Schlatters Disease), and the foot (Severs Disease, Kohlers Disease, Freibergs Disease). Avascular necrosis is the term used to describe a bone injury related to a loss of blood supply. Perthes Disease is similar in that the blood vessels in the femoral head are damaged, but Perthes Disease is different in that there seems to be repetitive small injuries rather that the catastrophic injury which occurs when adults get avascular necrosis of the femoral head.
DIAGNOSIS
The most common complaint leading to the diagnosis is a mild limp. Sometimes there is mild activity related pain in the hip or anterior thigh. Examination of the hip often demonstrates limited internal rotation. Xrays usually confirms the diagnosis, when there is increased density or collapse within the femoral head. If the condition has not yet reached that point, the xrays may look normal and a bone scan or MRI may be needed to make the diagnosis.

PERTHES DISEASE PROCESS
Perthes Disease is a process which reliably moves through several phases, which are divided into the initial phase, the increased density phase, the collapsing or fragmentation phase, the healing or reossification phase, and the final remodeling phase. These phases can be watched on xrays. Each phase is described below...

Initial Phase: Patients have a mild limp and maybe some achy pain with activity. Xrays may be normal or may show subtle changes with subtle lateralization or the femoral head ossification may be smaller than the other side.

Increased Density Phase: This phase lasts 6 to 12 months. Xrays show an increased density within the femoral head, but it is still round. There may be some changes (cysts) in the upper part of the femur.

Collapsing or Fragmentation Phase: This phase can last 6 to 12 months. The xrays show flattening and collapse of the dense bone. The head of the femur can slide laterally (subluxate) if there is a lot of collapse, especially when there is inflammation (often with achy pain) in the hip leading to muscle tightness (spasm) and limp. Subluxation is problematic and needs to be avoided by controlling symptoms and maintaining muscle flexibility. If subluxation develops and becomes fixed, it needs to be treated more aggressively with therapy, bracing, or surgery.

Healing or Reossification Phase: This phase can last 1-2 years or to maturity. Usually, there is no further worsening of the head and the size, shape, and density gradually improve. Once the body has removed all of the dense damaged bone, the healing process fills in new bone and the head gradually regains roundness. The femoral head must stay centered in the socket to help it heal in with good roundness.

Remodeling or Residual Phase: This phase is the final result obtained at maturity. The xrays show no changes in density or shape, but the internal structure of the bone continues to evolve over time. The final shape of the head is described with the Stuhlberg classification, which describes the roundness of the femoral head and its congruity within the socket.

TREATMENT PRINCIPLES
The goal of treatment is to get a painless, well functioning hip that does not wear out and develop arthritis over the next 50 years. Many different treatments have been used over the last 50 years. Treatments have included strict bedrest, bedrest with traction, body casting, bracing
without weight bearing, ambulatory bracing, various types of surgery on the femur and pelvis. Scientists have analyzed the results of these treatments, but it is not an easy problem to study. Studies that look at painless, well functioning hips show that essentially all the kids are painless with well functioning hips by the time they are 16-18 years old. In other words, essentially all patients with Perthes disease do very well in the short term once they reach maturity. This is irregardless of how they were treated.

Other studies have looked rates of arthritis 50 years out from treatment and it seems that hips that healed round do better than hips that healed flat. Stuhlberg class I-II hips that are round tend to hold up very well over time. Stuhlberg class III-IV hips with mild flattening hold up pretty well for 40-50 years. Stuhlberg class V hips are very flat and seem to wear out with osteoarthritis after 30-40 years. Hips that do wear out after 30 or more years do quite well with hip replacements.

Studies that have used roundness as the goal (as a predictor of rates for osteoarthritis 30 to 50 years from now) seem to suggest that it is important to keep the femoral head centered in the socket. The motion of the hip is a good guide to whether or not the femoral head is centered in the socket. The hip motion is decreased if there is pain and spasm, or if there lateral collapse and subluxation. Lateral collapse and subluxation is more common in children over age 8 and surgery is often recommended to prevent it. The surgery attempts to put the femoral head deep in the socket or to move the socket out over the femoral head. Keeping the femoral head deep in the socket is felt to help mold the femoral head and maintain its roundness. This is called containment.

There is no one right treatment for Perthes Disease. Important components of treatment include pain control, maintenance of motion, quality of life, and final shape of the femoral head. Pain is usually well controlled by adjusting activity and with simple pain medications like acetaminophen (Tylenol) or ibuprofen (Motrin, Advil, Etc). Motion should be monitored by the family and used as a guide to treatment. Usually, if pain is controlled, then motion is also well maintained. Quality of life is not unimportant. Running, jumping, biking, and playing sports are important in the lives of children. While these activities may worsen the Perthes disease process, it is excessive to completely prohibit these activities for the long period of time it takes for the Perthes disease process to run its course. Generally, limited participation in these activities is allowed, provided pain is pretty well controlled and motion is maintained. The final shape of the femoral head is related to patient age and keeping the femoral head in the socket. The motion of the hip is a good guide to whether or not the femoral head is centered in the socket. The goal of treatment is to get a painless, well functioning hip that does not wear out and develop arthritis.

**TREATMENT**

The mainstay of treatment for most children is focused on controlling symptoms and maintaining motion. Symptoms in Perthes disease are limping and pain related to activities. If symptoms are
present, they should be treated with activity restrictions and appropriate interventions for pain. If pain improves then pain interventions can be stopped and activities can be increased again.

It is very important to maintain motion in the hip. Hip motion is assessed by measuring flexion, extension, internal and external rotation, and abduction. Your doctor will show you how to measure these motions. If motion is lost and not regained after 2-3 weeks of anti-inflammatory pain medicine and activity restriction, then additional interventions may be needed including physical therapy, casting, surgery to lengthen tight tendons, or surgery to realign the hip joint. Parents should contact the doctor’s office if the child loses more than 25 degrees of motion and it does not respond to symptomatic treatment over 2-3 weeks.

Theoretical considerations related to the causes of Perthes disease can also be used to postulate interventions that could help to minimize damage and promote healing. Eating right and taking a daily vitamin are good for everybody. Getting regular cardiovascular exercise and avoiding caffeine and second hand smoke help to promote good blood flow and reduce spasms in blood vessels. Nutritional supplements such as condroitin sulfate and glucosamine may also help to promote cartilage healing.

**ACTIVITY RESTRICTIONS**

Running, jumping, biking, and playing sports are important in the lives of children. It is probably excessive and not needed to completely withdraw children with Perthes disease from all physical activity. However, it is appropriate to encourage low impact activities like swimming and biking over football and soccer. When motion is good and symptoms are mild or absent, limited participation in gym and sports should be allowed.

Parents are the best judge of appropriate activity restrictions. While it may be possible to figure out exactly how much time can be spent playing soccer or bike riding without becoming symptomatic, this may be difficult determine and even more difficult to enforce. My guidelines for activity modifications are to adjust activity by broad levels at no more or less than every 2 weeks. Activity levels can be set however the parents choose, but my recommendation is to consider four broad levels. Level one is full activity including gym and sports. Level two is no gym or sports, but running and jumping are allowed in play around the house. Level three is no running or jumping. Level four is required crutch walking. Decisions for setting the activity level are based on the family assessment of frequency of pain complaints, presence of a limp, cooperation with chores and schoolwork, and the motion in the hip. If these symptoms worsen, activity is decreased one level. If these symptoms are acceptable or improve, then activity is maintained or increased one level. Schools are required by federal law to make appropriate accommodations for children with medical conditions. You will need to notify school personnel of the child's medical condition and work out a system for keeping them informed regarding activity restrictions.

**PAIN MANAGEMENT**

Appropriate pain interventions include pain medications and any other interventions that help with the pain. The use of medications like acetaminophen (Tylenol) or ibuprofen (Motrin, Advil, Etc)
should be liberal as these medications are quite safe. The anti-inflammatory medications also help to reduce inflammation in the joint, which is probably of extra benefit in reducing muscle tightness. Anti-inflammatory are most commonly used for management of arthritis and we know form these patients, that (1) these medications don’t wear out or lose effectiveness (2) tolerance/addiction are not issues, (3) they work better if taken in advance of activities which might cause symptoms, and (4) they work even better if they are taken on the regular 2-3 times a day schedule. Taking these medications on a schedule avoids issues related to medications at school, as ibuprofen 3 times per day can be given at breakfast, after school, and at bedtime, while naprosyn 2 times per day can be given at bedtime or breakfast and dinnertime. Symptoms which arrive at school are best managed with rest or sitting, not medications. If needed, acetaminophen can be given in addition to the anti-inflammatory medications, and this may valuable as an extra tool to use when symptoms at school are difficult to manage.

Other interventions for pain include massage, stretching, ice, heating pads, and warm baths. Find out which interventions help your child and use them as needed.

**MONITORING HIP MOTION**

It is very important for the family to monitor hip motion, especially when symptoms are present. The hip is a ball and socket joint and can move in many directions. Five motions can be used to assess the hips function. Hip flexion and extension can be measured by bending one leg up to the chest, while keeping the other leg straight. It should be possible to bend one leg all the way to the chest (full flexion), while keeping the other leg out straight (full extension). Hip rotation is assessed by pointing the thigh and knee straight up, and checking to see how far the foot turns to the right and left. Finally, hip abduction is checked with the hip and knee extended and measuring how far the leg can be move outward. Hip motion should be measured and recorded at least once per week and whenever you are worried about the child’s symptoms.

**SURGICAL TREATMENT**

Surgical treatment is indicated for older children with whole head involvement and for younger children with lateral subluxation and a loss of motion that does not improve with symptomatic management. Osteotomy of the femur and/or pelvis attempt to put the femoral head into a contained position within the socket, allowing further growth and remodeling to occur in a round and congruous fashion. The osteotomy is usually fixed internally with screws and plates so that motion exercises may be started early.
**COMPLICATIONS**

Perthes disease is a process which always runs its course. It always heals eventually. Symptoms come and go during this process and should be managed appropriately. Problems along the way relate to pain, limp, and loss of motion. Management of these issues is described above. One usual complication that can develop is that a piece of cartilage can break free from the head and end up in the joint space and cause problems with motion. This is called osteochondritis dessicans. It is not known why it occurs. It usually affects older children with whole head involvement. It usually does not occur until late in the healing process. It is managed symptomatically and can improve on its own. Other times surgery is needed to remove the loose piece from the joint. The goal of treatment is to get a painless, well functioning hip that does not wear out and develop arthritis over the next 50 years. All children heal the Perthes disease, and the hip almost always functions well by the end of adolescence. However, in the long run, children who heal with flatten femoral heads have a higher than average risk of osteoarthritis after 30 to 50 years.

**PROGNOSIS**

Poor prognostic signs are age greater than 8 at presentation, involvement of the lateral column of the femoral head, poor range of motion, and lateral subluxation of the femoral head during healing. In almost all cases, symptoms resolve after the healing phase and the patients have complete freedom from pain during the teenage years and young adulthood. However, it is head of the femur is flat, it is estimated that approximately 50% will need hip replacement by later adulthood (50-60 years old). The Perthes Study Group has recently published result of their carefully designed long term study. This study compared treatments used at different centers to the final x-rays results at maturity. In general, kids less than 8 do best with symptomatic management. After age 8, results depend on the degree of lateral collapse. Mildly involved hips do best with symptomatic treatment. Moderately involved hips do best with surgery. Severely involved hips often heal with flattening irregardless of treatment choice.

**MORE INFORMATION**

Further information can be obtained on the internet. Your local public library can help you explore these sources if you are interested. Two good sites for expert and peer reviewed information are the American Academy of Orthopedic Surgeons at www.aaos.org and www.emedicine.com. Additional web sites are the Perthes Association at www.perthes.org.uk and the National Osteonecrosis Foundation at www.nonf.org.

**FEEDBACK**

If you have questions or comments, please contact the office or submit them to the web site at www.pedortho.com.