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Understanding Perthes

Perthes disease, also called Legg-Calvé-Perthes disease, is a condition in which the bone in the ball in a child's hip joint loses its circulation and becomes brittle. Over time, this causes the bone to die, but as circulation returns to the joint, new bone gradually grows in place of the brittle bone. For some children, this process affects the shape of the ball at the top of the thigh bone, which is called the femoral head. This can occur in one hip or both hips.

This stoppage of circulation in the hip can occur at any age, but the problem has different names for other age groups. A name that applies to any age group is avascular necrosis (AVN). Avascular means the loss of vascularity (blood flow) and necrosis means death. So, the term AVN means death of the bone because of loss of blood supply. However, doctors use a lot of different terms for this condition. You don't need to remember all of the names that are used for Perthes. They are explained here and are also in the glossary for reference. Perthes disease is the most common name, but you may hear Legg-Calvé-Perthes disease, or simply AVN because AVN is the medical term for what happened. In fact, AVN can occur in babies and in adults, but it's only called Perthes in the age group between 2 and 14 years. To make matters more confusing, AVN in babies and adults is also called osteonecrosis, or dead bone, in addition to being called AVN. So, you can see that doctors use many terms to identify one problem. This is similar to the way we identify ourselves by

our names, full names, nicknames, family relationships, or other terms.

One possible reason that the term Perthes is used for the age group between 4 and 13 is how Perthes affects the shape of the ball at the top of the thigh bone. Babies with AVN always recover with a round ball, but adults never do without treatment. The age between 4 and 13 is unpredictable. Some children in this age group recover without any intervention and others need treatment. So, the first question to ask is: who needs treatment and who doesn't in the age group between 4 and 13 years? Doctors rely on observations, tests, and experience to help decide who needs to be treated and who can be allowed to heal without intervention. We hope that this book will give you some insight into the keys for those decisions.

Hip Anatomy and Blood Supply

A hip joint is a ball-and-socket joint. The top of the thighbone (femur) is round, like a ball, and is called the femoral head. It fits deep inside the hip socket, which is called the acetabulum.

Cartilage covers the surfaces of the joint, and allows it to glide smoothly. Oily fluid inside the joint keeps it lubricated. That fluid is made by the lining of the joint called the synovium. A rim of soft tissue called the labrum surrounds the hip socket: it adds extra support and provides a seal that helps keep the ball inside the socket. Because it is not made of bone, the labrum cannot be seen on x-rays.

In Figure 1, notice that the blood vessels do not cross the growth plate because it is cartilage in the child. This limits the blood supply to the ball until the growth plate turns to bone as an adult. Sometimes Perthes causes the growth plate to turn to bone prematurely and this can cause a growth disturbance.

The blood supply to the hip in children is different from adults. Loss of blood supply leads to Avascular Necrosis (AVN) but that's the term that doctors use to say that the hip is crumbling because it has lost the blood supply—as opposed to damage

from trauma, infection, or other conditions. Some special aspects of the anatomy in children between the ages of five and ten years make circulation in the hip joint more precarious. When a child has Perthes, the small blood vessels that give blood to the ball become clotted and that stops the circulation. The ball of the hip does not have overlapping circulation as most other areas of the body, so the hip is especially at risk. Also, the blood vessels of the child's hip need to go around the growth plate and that makes children different from adults where the blood vessels freely travel up the middle of the bone.

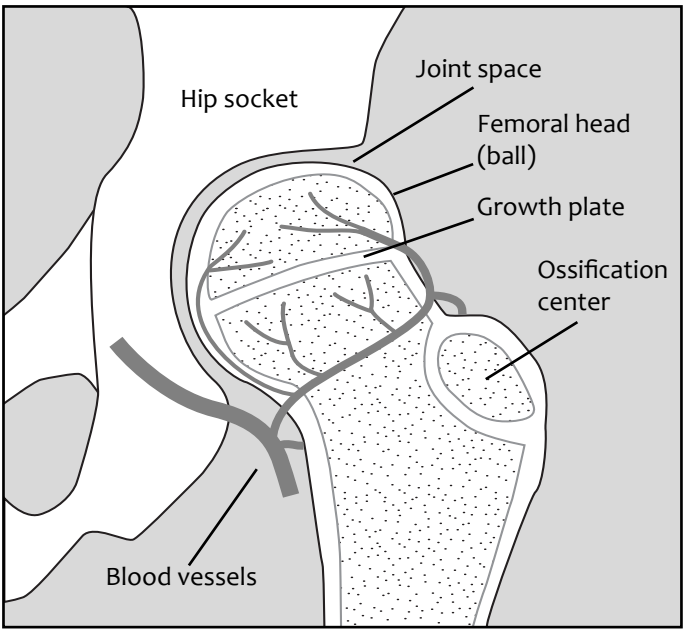

Figure 1. Normal hip anatomy for a child.

Range of Motion

The hip joint is a ball-and-socket joint like the shoulder, so it can move in all directions. When you walk, the hip joints flex forward and extend backward as you move your legs.[1] The knee also

[1] In this book, the word *leg* means the part of the body from the pelvis to the ankle. Doctors may use the term l*ower limb* to mean this part of the body, and use the word leg to mean only the part below the knee.

flexes and extends, but it is a hinge joint and can't rotate in all directions.

So why does the hip need to be a ball-and-socket joint? One reason is to allow the leg to swing out to the side (abduction), or inward toward the body (adduction). Actions like riding a horse or squatting involve abduction and adduction. The hip also rotates as you twist at the waist while walking, creating a longer stride. In track and field events, hurdlers twist their hips even more to get one leg stretched out over the hurdle while the other pushes from behind. Ball-and-socket motion is important for understanding Perthes because any limit of rotation or ability to move the hip away from the body causes a limp even though the hip might flex and extend perfectly.

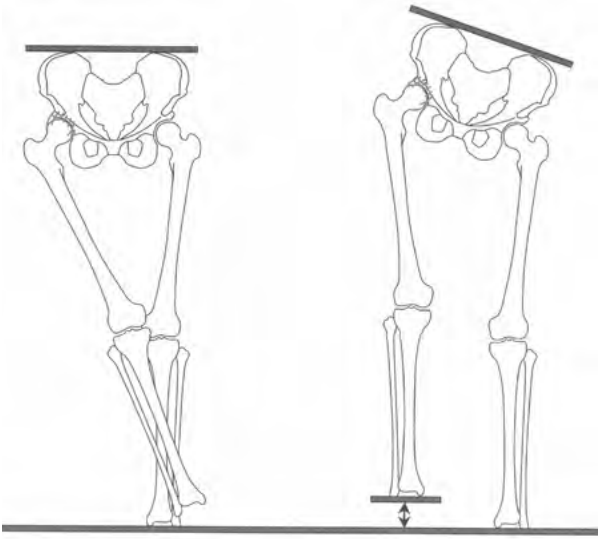

Figure 2. Crossing the legs with a painful limp.

If the ball in the hip joint becomes flat, the flattened ball limits the stride length, and causes the hip to jam when the pelvis tries to rotate.[2] The flattened ball also limits the ability of the leg to swing out to the side. The adductor muscles on the inside of

[2] Figure 2 reproduced with permission from D. Paley. Dror Paley, *Principles of Deformity Correction* (New York: Springer, 2002), 737.

the thigh may also be in spasm and prevent the leg from swinging to the normal position while walking or standing.

When the hip is painful, the leg may become stuck when crossing the midline because of muscle spasms or flatness of the ball. Then the child must hike the pelvis to walk and this makes the leg seem shorter than it really is.

Risk Factors and Possible Causes

Perthes is an uncommon disorder that affects more boys than girls in a ratio of five boys for every girl. While girls are less likely to get Perthes, they often have worse cases, or more long-term consequences. One reason is that girls stop growing before boys do, so there isn't as much time for the hip to recover completely. We don't know all the reasons for these differences between boys and girls although the increased activity level of boys has been considered as a possible cause. Perthes is also more common in low-birth weight children and in children with delayed maturity. This means that children with Perthes are often smaller than other children their age. Some children are exceptions to these trends, and that makes it difficult for doctors to be certain about what increases the risk of Perthes. Although several other possible causes of Perthes have been considered, there has been no conclusive evidence that Perthes is caused by parental cigarette smoking, increased blood coagulation, nutritional deficiencies, Attention Deficit Hyperactivity Disorder (ADHD), or kidney abnormalities. Researchers continue to look for causes.

The frequency of Perthes is difficult to estimate because ethnicity and the region in which a child lives seems to affect how common Perthes is even within the same country. About one in 10,000 white children in the USA will get Perthes, but the frequency in Black children is about 1 in 200,000. Asians have a rate of about 1 per 25,000, and Hispanics approximately 1 in 50,000.

Perthes affects children between the ages of 2 and 14 years old, and the peak age at onset is 5 to 7 years. Even the age at onset varies in some regions, and the peak age is older in India

than in Europe or the USA. It's not known whether the number of cases of Perthes is increasing or decreasing, but there is evidence in England that the total number of cases is decreasing, perhaps because of improved standards of living over the past fifty years.

Diagnosing Perthes

A diagnosis of Perthes has two parts. The first part is to make sure that the cause of the limp or pain is Perthes and not something else like an infection or an injury. The x-ray findings of Perthes are distinct, so a knowledgeable doctor can usually make the diagnosis based on x-rays. The second part is diagnosing the extent and stage. The extent means the amount of involvement or how much of the ball at the top of the thigh bone is affected. Younger children can tolerate more involvement than older children. Perthes goes through a series of stages. Chapter 2 describes each stage of Perthes and also discusses how the amount of involvement affects treatment.

The extent is hard to determine early in the course of Perthes when treatment decisions need to be made. Taking x-rays periodically helps because the extent becomes clearer as the bone changes. However, observing and waiting too long to intervene may decrease the chances of a good outcome. This is a major dilemma of Perthes: early treatment gives the best results, but it's not always easy to know who needs treatment in early stages.

To find out the stage, doctors use the child's history, physical exams, and x-rays. The history tells the doctor how long the child has been in pain or limping. Limping is a way to avoid pain, so doctors consider limping to be a sign of pain avoidance. Pain or limping for less than six months usually means the disease is still in the early stages.

X-Rays

In the early stages of Perthes, the x-rays may show few changes except for a crack under the joint surface, some increased white-

ness of the bone, or a slight change in the shape of the ball. Doctors use x-rays to monitor Perthes as it progresses. After a few months, the ball in the hip joint shows a moth-eaten appearance on x-rays as the dead bone is absorbed so that new bone can grow. In the later stages of Perthes, the ball is soft and may start to fragment or flatten and slip out of the joint sideways. The length of time the child has had pain or a limp plus the changes on the x-rays help determine the stage of Perthes.

It might seem that nuclear bone scans, CT scans, or standard MRI scans could reveal the extent of Perthes, but these methods tend to overestimate how much of the joint is affected. Plain x-rays are still the best method for estimating the extent of involvement of the ball, but this is an area of active research.

Concerns About X-Ray Hazards

X-rays are the rays that generate the image similar to the way that light rays generate an image in a digital camera or on photographic paper. Pictures of friends and family are called photographs. The images produced by x-rays are properly called radiographs, but we call these images x-rays even though the x-rays are the beams of energy that caused the images to appear.

In years gone by, lots of x-ray energy was required to produce the images on plastic films. Today's images are produced digitally with computer enhancement so the amount of x-rays needed to produce a good radiograph is very small. The same changes have occurred in photography, where today's digital cameras—especially video cameras—can take good pictures indoors without much light. Similar technology enhances the x-ray images.

The amount of x-ray exposure with digital imaging is so low that it is usually best to avoid shielding of the genital organs so that the best x-rays can be obtained on the first set of x-rays (radiographs). This is safe and standard in most offices or hospitals where children are treated. If you are in doubt, it doesn't hurt to ask the doctor if his x-ray equipment is digital or if he is using "rare earth" x-ray exposures. Either of those has very low irradiation. CT scans have a lot of x-ray exposure but MRI has none.

In the past, shielding was recommended because ovaries and testes are sensitive to radiation. A recent study[3] of pelvic x-rays of children found that the shields were placed incorrectly most of the time, and that the radiation exposure with modern x-ray equipment is very small. The study concluded that it might be better to stop the shielding.

Figure 3. Early Perthes. The hip on your right has Perthes.

Figure 4. Later Perthes in the same child as Figure 3.

Interpreting X-Rays

X-rays taken in the early course of Perthes may only show that one hip is whiter and a little flatter than the normal hip. Initially,

[3] Marij J. Frantzen et al., "Gonad Shielding in Paediatric Pelvic Radiography: Disadvantages Prevail Over Benefit", Insights into Imaging 3.1 (2012): 23, http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3292647/.

it is hard to detect this difference but the whiteness becomes more pronounced after a few weeks. The whiteness is because the blood stops circulating, and fluids that are rich in calcium seep into the spongy bone. Limping also leads to bone loss around the dead bone and this makes the contrast more apparent. The area of white bone is the first clue to the extent of Perthes. When this is clearly seen, it usually shows how much of the ball is affected. A small central area of whiteness is less worrisome than complete involvement of the ball where the whole ball is white. (The anatomy of a child's hip is shown in "Figure 1. Normal hip anatomy for a child." on page 3.)

Sometimes a crack below the surface of the joint is visible where the brittle dead bone has broken. This break usually causes some pain for the child and may be the first sign of Perthes. The length and location of the crack has been used to classify the severity of Perthes with the Salter-Thompson method. (See "Perthes Classifications" on page 37.)

Unfortunately, it's not a reliable method because the x-rays rarely show the whole crack, and may not show the crack at all. Imagine shining a light through a crack in a wall. If the light isn't lined up perfectly with the crack, then the light won't shine through to the other side. If the x-ray beams don't line up with the crack in the ball then the x-ray beam won't shine through onto the radiograph.

Interpreting x-rays of Perthes is more about what you don't see than what you do see, because the changes visible on x-ray lag behind the process by four to six months. As time goes by, the white bone is taken away by blood vessels that grow into the ball to help it recover. Then the ball begins to look moth-eaten because the blood vessels don't show up on the x-rays. The areas of bone that look like they are gone have been replaced by blood vessels and cartilage as part of the healing process. The ball becomes soft and likely to collapse where the white bone is disappearing during this stage. If only a small part of the ball is affected, then the normal areas will give support and keep the ball from collapsing.

Another area that doesn't show up on the x-ray is the joint space. This is the area between the ball and the socket, but it's not actually a space because it is filled with cartilage on the surface of the ball and the lining of the socket. The bone has blood, but the cartilage surface does not. When the bone loses blood supply, the bone becomes brittle and no longer supports the cartilage surface.

If the ball collapses, it becomes very white and thin and the ball does not take up as much space in the socket. This allows the ball to move upward and slide out of the socket sideways. Some x-ray measurements can help determine if this is happening. Perhaps the best way is to look at something called Shenton's line.

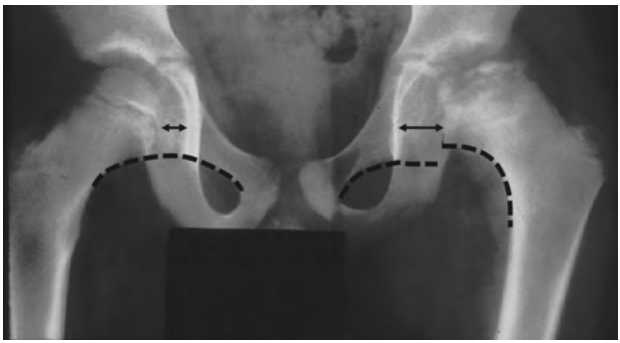

Figure 5. Shenton's line is broken.

Shenton's line is the smooth arc across the bottom of the hip joint where the ball is in the socket. Figure 5 shows a broken Shenton's line on the side with Perthes.

Arthrogram (X-Ray with Dye)

Arthrography, also called an arthrogram, is an x-ray taken after dye is injected into a joint to show more detail than can be seen in a plain x-ray. Anesthesia is used during this procedure. The hip is always round at the beginning of Perthes, so arthrography has little role during the early stages. An arthrogram is generally used if the hip has begun to slide out of the socket sideways to see if the hip can be put back into the socket.

The arthrogram also helps show whether the ball is too flat to recover with standard surgery or by putting the child in a cast for an extended period of time. An MRI may show the flatness of the femoral head, but general anesthesia during arthroscopy relaxes the muscle spasms and lets the doctor determine if the hip is too far out of the socket to go back in. If the hip cannot be put back into the socket, then salvage procedures may be needed.

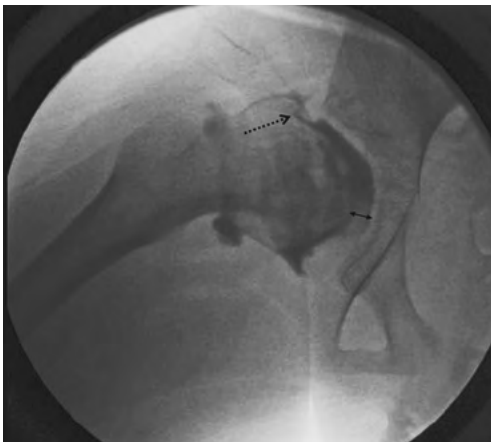

Figure 6. Hip arthrogram.

In Figure 6, the dye in the joint is black and the dotted arrow points to a dent in the ball that is hooked on the edge of the socket. The dye shows up between the ball and the edge of the socket because the ball cannot be put back into the socket completely. When Perthes has reached this stage, doctors call it "hinge abduction" and containment is rarely possible or successful. (See "Hinge Abduction" on page 20.)

Computed Tomography (CT)

Computed tomography (CT) is commonly called a CAT scan. This is rarely needed for children with Perthes, and it exposes the child to many more x-rays than other types of imaging. Newer CT machines have less radiation, but it's worthwhile to question your doctor about alternatives whenever a CT is recommended.

Current CT technology does not accurately show how much of the ball is affected by Perthes. During a CT, the x-ray beam moves around the body so that images can be seen from many angles. The final images show "slices" of the area being scanned.

Magnetic Resonance Imaging (MRI)

In Perthes, the standard MRI may be helpful to diagnose Perthes. MRI does not use x-rays. It uses a strong magnetic field and radio waves to create images of tissues. This allows the doctor see the ligaments, muscles, and tendons (also called soft tissue) around a joint much more clearly than on an x-ray. An MRI lets the doctor see the joint surface and the joint shape. It's necessary to stay very still for 15–20 minutes while the MRI is being done, so babies and children usually need sedation or general anesthesia for an MRI so that they stay still and the images are clear.

Standard MRI is not very helpful to guide treatment. This is because the standard MRI tends to overestimate or underestimate the extent of the damage. However, perfusion MRI, also called contrast MRI, is more sensitive in detecting how much of the femoral head has lost its blood flow. In perfusion MRI, a contrast solution is given through a vein before obtaining the MRI. Since the contrast goes to the area of the femoral head that has blood flow, it shows the area without blood flow as a dark region.

One shortcoming of MRI is that the hip cannot be moved around like it can during an arthrogram (x-ray with dye), so an arthrogram is usually preferred in later stages of Perthes to determine whether the ball can be contained in the socket. However, in some situations the MRI can help define the shape of the ball if the child can hold still without general anesthesia.

The doctor might recommend an MRI with dye injected into the hip joint. This is called an MR arthrogram and is very rarely used for Perthes. An MR arthrogram may be needed later if a child has pain after the Perthes has run its course and the hip is not round. In these cases the soft tissues around the joint may

be damaged and imaging may need dye in the joint to enhance the MRI. For instance, a teen who is thought to have a tear in the labrum (the soft rim of cartilage that surrounds the hip joint) might need an MR arthrogram. Sometimes a high-resolution MRI machine can reveal more detail about the joint and avoid the dye injection, but not all facilities have the highest resolution machines. If dye is used, a local anesthetic is injected into the hip joint first, then the contrast material is injected.