



Perthes Disease



The Parents' Guide





Introduction

At Steps, we understand how a lower limb condition can affect individuals, families, and communities. Our commitment is to helping people understand these conditions, offering reassurance and actively working for a better future, through our work with national health services and research projects.

This booklet is for parents with a child diagnosed with Perthes Disease. It cannot tell you everything you need to know about what the future may hold, but we hope it will reassure you and your child. It is also intended to show that practical help, specialist medical information, emotional support, and links to other sources of information are all available, if needed. This will help you to be more prepared for the road ahead and have information to hand so that you can ask informed questions about your child's care, treatment and prognosis.

Help when you need it

Sometimes being able to contact someone who knows what you are going through can provide much needed encouragement. Our Family Contact Service can put you in touch with others who have shared a similar experience and can offer advice, support, and practical tips. You can also share your problems and solutions to everyday challenges on our closed Facebook Group for parents. The group is a friendly and safe way of discussing online your worries with other parents, sharing tips, and finding emotional support.

Remember, the STEPS Helpline team are here to offer information and support in total confidence and answer any questions or concerns you may have. This will help you to ask informed questions at hospital appointments or may help to reassure you along the way.

No matter how big or small your concern, please telephone our Helpline on **01925 750271** or email info@steps-charity.org.uk for support and advice in total confidence. Social media details can be found on the back cover.



Testimonial

Declan and Gemma's Story

"My name is Gemma and my son had Perthes as a child. When he was 7, I took him to the doctor as he was in agony with a lot of pain in his hips and in his knees. He had aches and pains in his legs, and he was limping. At first there was some confusion as to the cause of the pains, it was thought that he might have fluid on his hips. However, after discussions and examinations by another consultant he was diagnosed with Perthes Disease.



Declan Thompson -
(Young ambassador for Steps Charity
Worldwide and professional footballer
for Sheffield Wednesday)

My son does not remember much about the treatment which started when he was 8 years old, we had waited for almost a year from diagnosis to surgery. For a year before the surgery he was in a wheelchair and had to be carried upstairs. The treatment involved an operation in Sheffield Children's Hospital, an osteotomy. This surgery was considered necessary to improve the position of the ball and socket in the hip joint and to protect it from losing the round shape. After the operation he used crutches for around 6 weeks but eventually he was able to discard the crutches and hop around. He also tried to carry on playing football with his crutches in hand; there was no stopping him.

In the weeks and months after the operation my son attended physiotherapy, this was a great help to him as it relieved the pain and helped him to strengthen his muscles. The weekly physiotherapy sessions were not just helpful in regaining his physical strength, but they helped build his confidence and he soon became determined to get better and get on with his life.

Testimonial . . .

As part of his recovery he went swimming, this helped a lot as it was great exercise and enjoyable. He attended hospital for reviews and regular check-ups, and all the time he was building his strength and was playing football with good mobility.

“My son has overcome his challenges, he is a healthy, active, motivated, 18-year-old and is a professional athlete”.

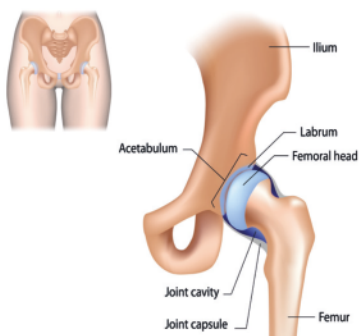
When we first got the diagnosis of Perthes we did not know anything about the condition, since then we have met and know many families where children have been treated for Perthes. We are happy to share our story with Steps as there will be children and parents who need to have information about the condition.

Parents and children need to know that with Perthes there are a range of treatments that can help, indeed many children do well without any surgical intervention and often the hip regrows into a good shape. Early diagnosis is important as Perthes can be managed and it helps avoid problems later in life. The Steps website and social media channels have lots of information on Perthes including videos, webinars with orthopaedic consultants, family stories and factsheets.”



What is Perthes Disease?

Perthes disease, or Legg-Calve-Perthes disease as it is sometimes known, is a condition which develops in childhood and affects the growth of the hip joint. There are two parts to the hip joint: the ball (the femoral head) and, the socket, (the acetabulum). The structure of the hip is shown in the diagram below.



In cases of Perthes, the blood supply to the ball part of the joint becomes disrupted. This means that no blood gets to the femoral head (necrosis), affecting the way the joint grows and causing the bone to flatten and become squashed. This, in turn, affects the hip shape and the way it works.

Why does it happen?

It is not known exactly why the blood supply to the hip becomes affected. Studies have proven that it could be an exposure to something within the environment which triggers the Perthes Disease process. In young children there is only one blood vessel which runs to the top of the hip joint. It is thought that this blood vessel becomes blocked off. The hard bone within the hip becomes unhealthy and the hip softens.

Perthes Disease is not thought to be caused by physical injury or a problem with blood vessels in the rest of the body. There are no childhood conditions linked to Perthes Disease. It is rare and occurs in less than one in 1,200 children. Interestingly, we know it happens in Northern parts of the UK more than anywhere else in the world. There is evidence that Perthes Disease is not genetic. However, interestingly, we know that some families have more than one family member affected.



How long will recovery take?

In most cases, over time, the affected blood vessels begin to regrow, allowing new bone to be produced in the unhealthy areas. Recovery usually takes a few years as the bone around the femoral head reforms – in a similar, but more prolonged way, to the recovery of a bone following a fracture.

Symptoms and Diagnosis

Perthes is approximately four times more common in boys than girls. Most cases occur between 4 and 8 years of age, and most occur in Caucasian (white) children.

Affected children are often amongst the smallest in the class, but it is not fully understood why. The condition mainly happens in one hip (unilateral) but in 1 in 10 children, it can happen in both hips (bilateral).

The first symptom is usually a limp. Pain may or may not be a feature, and similarly stiffness of the joint may or may not occur. Often pain is sometimes only felt at the knee

(known as referred pain). This is because the nerve from the hip travels on to the knee and can transfer the pain signals downwards. Sometimes, pain may build up over a few weeks.

An x-ray will usually confirm a diagnosis of Perthes. Sometimes blood tests are performed to rule out an infection of the bone and check for inflammation. Special tests (like an MRI or bone scan) may also be used to help make the diagnosis, though, these tests rarely provide any more useful information and are usually of little help if the initial x-ray clearly shows Perthes Disease.

It is possible that, if a joint infection is suspected, a small sample of fluid is taken from the hip. This is usually done with the child asleep under a general anaesthetic.



Stages of Perthes Disease

Perthes Disease goes through a number of different stages as it heals.

A – Initial (necrosis) stage – In this stage of the disease, the blood supply to the femoral head is disrupted and bone cells become unhealthy. On x-ray, the femoral head may look normal, or may have very subtle changes compared with the unaffected hip. The hip becomes inflamed and your child may begin to have symptoms, such as a limp or different way of walking. Sometimes it may be difficult to identify that a child has Perthes Disease as the x-ray appears normal and an MRI may be considered.

B – Sclerosis – This stage is called sclerosis as the top of the femur bone becomes very white as the bone structure changes. It is during this phase that the bone is in a weaker state and the head of the femur is more likely to collapse into a flatter position, called the Coxa Plana.

C – Fragmentation – In this stage new blood vessels have formed to help the hip reform. The new blood supply removes the unhealthy very white bone and begins to replace it with healthy bone. On an x-ray, the

femoral head looks 'patchy' with white and dark areas – as unhealthy bone is removed, and new bone begins to be formed.

D – Re-ossification – This is the stage when the new bone formation occurs most rapidly. The unhealthy bone has been removed. The new bone grows from the outer perimeter of the femoral head and gradually fills the central area. The re-ossification stage is often the longest stage of the disease and can last a few years, typically 2 to 3 years. The end result of the re-ossification stage is the complete bone regrowth with the femoral head reaching its final shape. The 'rounder' the hip, the better the result. How close the shape is to round depends on the extent of damage in the fragmentation phase, the child's age at the onset of disease, with younger children (particularly under 5 years old) usually having the best results.

The final shape of the femoral head and how it fits the socket will determine the long-term outcome.



Treatment

The hip in Perthes Disease **always** regrows – though the shape of the hip is the reason that doctors monitor the hip closely. The ideal hip is a football shape – as these hips work normally. The next best hip is a rugby ball shape – as these work well, though may wear out early. The hips that cause problems, and the most pain, are very flat, or odd shaped – as these don't tend to move normally.

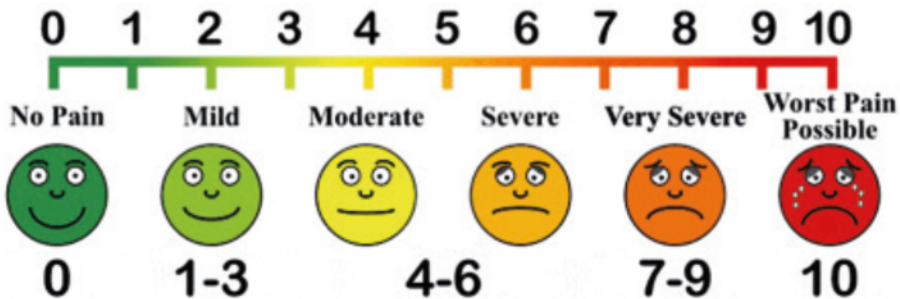
Many children with Perthes Disease do well without an operation and the hip can regrow into a good or at least reasonably well working shape.

However, it is hard to identify the children with the condition who are likely to do well if they have an operation, and those who will do just as well if they do not have surgery. Medical agreement on the best way to treat Perthes has not yet been reached, although study into this is ongoing.

provided with a pain chart so your child can express how much they are hurting consistently.

Physiotherapy and/or hydrotherapy are sometimes used to help with the movement of the joint and to relieve pain. Sometimes, surgery to the femoral head (top of the thigh bone) or socket is performed.

Any surgery will be done with the aim of keeping the ball-shaped head of the thigh bone in the hip socket and improving the movement of the joint. This will help preserve the shape of the head of the femur (the ball) and keep it as round as possible. The more round the femoral head is when the disease has passed, the better the long-term function of the hip and the better a child's movement will be.



Pain relief, such as analgesics (pain killers) and anti-inflammatory medicines can help relieve the worst of the symptoms. Restricting high impact activity is commonly advised to alleviate symptoms and prevent further worsening of the problem. You may be

Swimming is often recommended as an activity to help with the movement of the joint because it is a good, low impact way of keeping the hip moving, but high impact exercise such as jumping may be restricted.

Possible treatment methods include:

Observation - if the hip is positioned well, regular outpatient appointments may be all that are needed to monitor the hip joint and range of movement as it re-shapes itself. Regular x-ray monitoring will confirm this.

Traction – if the condition is associated with pain and/or poor range of movement, traction may occasionally be used. Traction uses bandages and weights to ease the pressure on the joint and blood supply.

Surgery - may be considered to improve the position and containment of the ball into the socket and protect it from losing its round shape.



Surgical options

There are many different surgical options for Perthes Disease, this is because the condition is variable and, there is some degree of uncertainty amongst surgeons. Any treatment will be tailored to your child and considers factors such as their age, size, weight, severity, and progression of the disease. Two children, treated by the same specialist, in the same hospital, may have very different treatments and receive different advice on suitable activities and recovery times.

Some of the surgical options for treating Perthes Disease are:

Soft tissue release - release of tight muscles in the groin that reduce the range of movement in the hip.

Femoral osteotomy - an operation to alter the position of the 'ball' part of the joint at the top of the femur.

Pelvic osteotomy - an operation to alter the position of the hip socket.

Shelf acetabuloplasty - enlarging the hip socket using bone from elsewhere on the pelvis.

Some surgeons may use a hip spica cast after some of the above operations.

The final aims of such treatments are to help your child have a normally formed hip joint when they grow up and avoid the development of 'wear and tear' arthritis in adult life. This will help your child be as pain free as possible when they are adults.

What is a hip spica cast?

A hip spica cast is a large plaster cast that can best be described as a 'plaster of paris' pair of trousers. Traditional plaster of paris may be used over wadding, or a combination of plaster of paris and fibreglass material or all fibreglass. Plaster of paris is always white, but, depending on the hospital, the fibreglass plasters can be coloured or even patterned.



The shape of the spica varies and can extend from the mid-chest down to the ankle, sometimes with a bar across. If the problem is only on one side, the cast will probably extend to the ankle on the affected side and may stop just above the knee on the unaffected side, but this depends upon the hip stability which will be judged by the surgeon. A 'letterbox' style hole is left in the groin area to allow for toileting. The purpose of a hip spica is to keep the affected hip in the best position in order for it to recover from surgery.

What about activities?

Most doctors suggest the avoidance of high impact activities (i.e. jumping, contact sports) whilst the bone is 'soft'. As the bone hardens, children are permitted to go back to their usual activities.

Long term outlook



The long-term outcome depends on the final shape of the hip joint at the end of childhood; if the hip is reasonably round at that stage, the long-term outlook is usually good.

In some cases, despite treatment, the hip will remain very flattened or abnormally shaped. In these cases, 'wear and tear' arthritis is likely to develop in relatively young adulthood, making a hip replacement necessary at a young age. About half of children with the condition will be vulnerable to arthritis in later life and may require an 'early' hip replacement (in their fifties).

In all children with Perthes Disease, the blood supply to the hip will improve overtime, however they are treated. The hip continues to grow and reshape over time. Children under five, when they are diagnosed, usually have a very good chance of a full recovery, because of the longer time the hip has to rebuild itself as the child grows.



Personal Stories

Fred's Story



When Fred was 3, he started complaining of pains in his left leg. At first, we thought he'd pulled a muscle. He was a typical 3-year-old boy running and jumping off

everything. He kept complaining, sometimes of pain in his left knee and sometimes he had a limp. We had several trips to the doctors, drop-in and Alder Hey. After about 3 months of being mis-diagnosed with a hip infection, irritable hip, he was diagnosed with Perthes Disease.

We were told it was so severe he would need an operation straight away as his movement was so bad. They put us on the waiting list at Alder Hey and told us to come back in 6 weeks for his pre-op. His pain went from bad to worse very quickly. He didn't sleep and sometimes he couldn't put his left foot down with the pain. After a few trips to Alder Hey we were given strong meds to give him at home, but these would just knock him out: although he needed the pain relief it was horrendous to see him like this.

We went for his pre-op to be told they didn't want to do the operation on someone so young so for now; after a

second opinion at Alder Hey we were told we just had to see what happens. We were heartbroken not knowing what was going to happen, what we could do or how long this would go on for. Fred's doctor has been amazing and with us every step of the way. The support from him has gotten us through. The doctor monitored him closely and Fred has never had an intervention.

It had a huge impact on our lives in the early days, we had to totally change the way we lived our lives; we were limited to what we could do and where we could go. Simple things like going for a walk, to parties, who we visit, where we go, how much travelling etc. We had to consider how much walking was involved. There were very few parties he could go to as they were normally soft play, football, or trampoline parks.





A success story

Fred's Story . . .

We had always been an active family, and this had changed in terms of us being active together. We couldn't just go to the beach for a walk with the dog. Fred could walk but not for any length of time as his leg would get sore. Generally, we used a pram, but he hated going in it. He would as long as we explained to people about his hip and why he was in a pram. His dad would put him on his shoulders but the position of his hip when he'd been on his dad's shoulders meant his hip would stiffen really quickly and get sore.

For myself and his dad it was just heart-breaking seeing your baby in so much pain. It was the pain that affected us the most: knowing as parents your child is screaming in pain and there's nothing you can do to help. You feel it in the pit of your stomach. It wasn't going to beat us, and we'd do everything in our power to help him and we have.

We are so lucky to have support from our amazing family & friends and Lily has always been the best big sister anyone could ask for.

Looking back now I think people just didn't understand, it's a rare disease and the other problem was on good days Fred looked like there was nothing wrong and he could do anything any other child could, but it was the fall out afterwards that his body couldn't cope with and he would suffer terrible if he did too much. People didn't see the bad side though. Very few people have.

We only put positive pictures on social media and hid away with the curtains closed when he had a bad day until he'd recovered. We would try to shelter him from people seeing him. We certainly didn't show how bad it got so I suppose it's hard for people to understand without seeing it.



A success story

Fred's Story . . .

Fred is now 7 1/2 and doing amazingly well. The blood supply returned to his hip about 18 months ago, he now has significant regrowth (nearly 50%) and Fred's doctor is really happy with the shape of the regrowth too.

To the naked eye it's hard to tell which side has Perthes Disease and that's down to hours of physio, massage, stretching and exercises. We've worked as a team since day one. I've done his daily exercises, soft tissue release, massage and physio. His dad has worked on his strength with training, teaching him to ride a bike (something he'd never been able to do in case he fell on his hip) he now goes on over 20km bike rides with his dad without complaining of any pain. He also plays for Marshall's, a local football team in Liverpool (again, something he'd never been able to do and was so desperate to play). His dad takes him training on Wednesday evenings and he plays matches every Saturday and he's loving every minute of being on the pitch!



The only other thing I'd like to say is Fred himself is amazing. He doesn't let anything hold him back. Of course, he still has bad days, but they are few and far between.

We're so ridiculously proud of him with how he's dealt with it and we'll always be grateful to our amazing family, friends and Fred's doctor that have got us through.

*"Fred's our hero,
our Perthes warrior."*

Further reading & references

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We don't take walking for granted...



Babies & Children



Anyone



Families

Where can I find out more information about Perthes Disease?

You should contact your medical practitioner for information relating directly to your child or for more general information contact STEPS.

www.steps-charity.org.uk

Helpline: +44 (0) 1925 750 271

Email: info@steps-charity.org.uk



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Helpline number

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email : info@steps-charity.org.uk

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