www.printo.it/pediatric-rheumatology/IE/intro

Limb Pain Syndromes

Version of 2016

1. Introduction

Many paediatric diseases can cause limb pain. The name Limb Pain Syndrome is a generic term for a group of different medical conditions with quite different causes and clinical presentations that share the presence of continuous or intermittent limb pain. To give this diagnosis, physicians perform investigations looking for known diseases, including severe ones that can cause limb pain.

2. Chronic Widespread Pain Syndrome (formerly called Juvenile Fibromyalgia Syndrome)

2.1 What is it?

Fibromyalgia belongs to the "amplified musculoskeletal pain syndrome" group. Fibromyalgia is a syndrome characterised by long-term widespread musculoskeletal pain involving the upper or lower extremities as well as the back, abdomen, chest, neck and/or jaw for at least 3 months, combined with fatigue, unrefreshing sleep and problems of variable intensity in attention level, problem solving, reasoning or memory.

2.2 How common is it?

Fibromyalgia occurs mainly in adults. In paediatrics, it is reported predominantly in adolescents, with a frequency of about 1%.
Females are more frequently affected than males. Children with this condition share many clinical characteristics with those affected by complex regional pain syndrome.

2.3 What are the typical clinical characteristics?

Patients complain of diffuse pain over limbs, although the severity of the pain may vary from child to child. The pain may affect any part of the body (upper and lower extremities, back, abdomen, chest, neck and jaw).

Children with this condition usually have sleep problems and the feeling of awakening from an unrefreshing, non-restorative sleep. Another main complaint is intense fatigue, accompanied by decreased physical capacity.

Patients with fibromyalgia frequently report headaches, limb swelling (there is a sensation of swelling although no swelling can be seen), numbness and, sometimes, bluish colour of the fingers. These symptoms cause anxiety, depression and many absences from school.

2.4 How is it diagnosed?

The history of generalized aching in at least 3 body areas, lasting more than 3 months, together with a variable degree of fatigue, unrefreshing sleep and cognitive symptoms (attention, learning, reasoning, memory, decision making and problem solving capacities) is diagnostic. Many patients present tender muscle points (trigger points) at certain locations although this finding is not required for diagnosis.

2.5 How can we treat it?

An important issue is to decrease the anxiety produced by this condition by explaining to the patients and their family that although the pain is severe and real, there is neither damage to the joints nor a serious physical disease.

The most important and effective approach is a progressive cardiovascular fitness training programme, and swimming is the best exercise. The second issue is to start a cognitive behavioural therapy, individually or in a group. Finally, some patients may need a drug therapy to improve sleep quality.

2.6 What is the prognosis?

Full recovery requires major efforts by the patient and essential support from the family. Generally, the outcome in children is much better than in adults and most of them will recover. Compliance with a regular physical exercise programme is very important. Psychological support, as well as medication for sleep, anxiety and depression may be indicated for adolescents.

3. Complex Regional Pain Syndrome Type 1 (Synonyms: Reflex Sympathetic Dystrophy, Localized Idiopathic Musculoskeletal Pain Syndrome)

3.1 What is it?

Extremely severe limb pain of unknown cause frequently associated with skin changes.

3.2 How common is it?

The frequency is unknown. It is more common in adolescents (average onset age is around 12 years) and in girls.

3.3 What are the main symptoms?

Usually, there is a long-term history of very intense limb pain that is unresponsive to different therapies and increases over time. Frequently, it results in the inability to use the affected limb.
Sensations that are painless to most people, such as light touch, may be extremely painful to affected children. This odd sensation is called "allodynia".
These symptoms interfere with the daily activities of affected children, who often miss many days at school.
Over time, a subset of children develops changes in skin colour (pallid or purple mottled appearance), temperature (usually reduced) or perspiration. Swelling of a limb may also be present. The child may sometimes keep the limb in unusual postures, refusing any movement.

3.4 How is it diagnosed?

Until a few years ago, these syndromes received different names, but

today physicians refer to them as complex regional pain syndromes. Different criteria are used for the diagnosis of the disease.
The diagnosis is clinical, based on the features of the pain (severe, prolonged, limiting activity, unresponsive to therapy, presence of allodynia) and the physical examination.
The combination of complaints and clinical findings is quite characteristic. The diagnosis requires that other diseases that generally can be managed by primary care doctors, clinicians or paediatricians are ruled out before referral to a paediatric rheumatologist. Laboratory studies are standard. An MRI may show non-specific alterations of the bone, joints and muscles.

3.5 How can we treat it?

The best approach is an intensive physical exercise therapy programme supervised by physical and occupational therapists, with or without psychotherapy. Other treatments have been used, alone or in combination, including antidepressants, biofeedback, transcutaneous electrical nerve stimulation and behavioural modification – all without definitive results. Analgesics (pain-killers) are usually ineffective. Research is currently underway and in the future better treatments will hopefully arise as the causes are identified. The treatment is hard for all people involved: the children, the family and the treating team. Psychological intervention is usually necessary because of the stress produced by the disease. Difficulty on the part of the family in accepting the diagnosis and in complying with treatment recommendations are the major causes of treatment failure.

3.6 What is the prognosis?

This disease has a better prognosis in children than in adults. In addition, most children recover faster than adults. However, it takes time and the lapse to recovery varies widely from child to child. Early diagnosis and intervention lead to a better prognosis.

3.7 What about everyday life?

Children should be encouraged to maintain physical activities, regular school attendance and leisure time with their peers.

4. Erythromelalgia

4.1 What is it?

It is also called "erythermalgia". The name for this condition derives from 3 Greek words: erythros (red), melos (limb) and algos (pain). It is extremely uncommon, although it may run in families. Most children are around 10 years of age when they start presenting complaints. It is more frequent in girls.

The complaints include a burning sensation with warm, red and swollen feet or, less commonly, hands. The symptoms worsen upon exposure to heat and are relieved by cooling the extremity, to the point that some children refuse to remove their feet from ice-cold water. The course is unrelenting. Avoiding heat and vigorous exercise seem the most useful control measures.

Many different drugs can be used in an attempt to relieve pain, including anti-inflammatory drugs, pain killers and drugs for improving blood circulation (called "vasodilators"); a physician will prescribe what is most appropriate for each child.

5. Growing Pains

5.1 What is it?

Growing pains is a benign term that refers to a characteristic pattern of pain in the limbs, which usually occurs in children between 3 and 10 years of age. It is also called "benign limb pain of childhood" or "recurrent nocturnal limb pains".

5.2. How common is it?

Growing pains is a common complaint in paediatrics. It presents with similar frequency in boys and girls, affecting 10-20% of children worldwide.

5.3 What are the main symptoms?

Pain appears mostly in the legs (shins, calves, thighs or in the back of

the knees) and it is usually bilateral. It appears late in the day or at night, often awakening the child. Parents commonly report that the pain occurs after physical activity.
Pain episodes usually last 10 to 30 minutes, although it might range from minutes to hours. Intensity can be mild or very severe. Growing pains are intermittent, with pain-free intervals lasting days to months. In some cases, the pain episodes may occur daily.

5.4 How is it diagnosed?

The characteristic pain pattern, combined with the absence of symptoms in the morning and with a normal physical examination, lead to the diagnosis. As a rule, results of laboratory studies and X-rays are always normal. However, X-rays may be required to exclude other pathologies.

5.5 How can we treat it?

Explaining the benign nature of the process reduces anxiety in the child and the family. During pain episodes, local massage, heat application and mild analgesics may help. In children with frequent episodes, an evening dose of ibuprofen may be helpful to control more severe pain episodes.

5.6 What is the prognosis?

Growing pains are not associated with any serious organic disease and usually resolve spontaneously in late childhood. In 100% of children, the pain disappears as they grow older.

6. Benign Hypermobility Syndrome

6.1 What is it?

Hypermobility refers to children who have flexible or loose joints. It is also called joint laxity. Some children may experience pain. Benign Hypermobility Syndrome (BHS) refers to children who present limb pain due to increased mobility (range of motion) of the joints, without any associated connective tissue disease. Therefore, BHS is not a disease

but rather a variation of the norm.

6.2 How common is it?

BHS is an extremely common condition in children and youngsters, present in 10 to 30% of children younger than 10 years of age and particularly in girls. Its frequency decreases with age. It frequently runs in families.

6.3 What are the main symptoms?

Hypermobility frequently results in intermittent, deep aching and recurrent pains at the end of the day or at night in the knees, feet and/or ankles. In children playing piano, violin, etc., it might affect the fingers instead. Physical activity and exercise may trigger or increase the pain. Rarely, mild joint swelling may be present.

6.4 How is it diagnosed?

The diagnosis is made on the basis of a pre-defined set of criteria that quantify joint mobility and the absence of other signs of connective tissue disease.

6.5 How can we treat it?

Treatment is very seldom necessary. If the child plays certain repetitive impact sports such as football or gymnastics, and develops recurrent sprains/torn joints, muscle strengthening and joint protection (elastic or supportive bandages, sleeves) should be used.

6.6 What about everyday life?

Hypermobility is a benign condition, which tends to resolve with age. Families should be aware that its main risk comes from preventing children from living normal lives.
Children should be encouraged to maintain a normal level of activity, including being involved in any sports in which they are interested.

7. Transient Synovitis

7.1 What is it?

Transient synovitis is a mild inflammation (small amount of fluid within the joint), of unknown cause, of the hip joint that resolves by itself leaving no damage.

7.2 How common is it?

It is the most common cause of hip pain in paediatrics. It affects 2 to 3% of children aged 3-10 years. It is more common in boys (one girl for every 3/4 boys).

7.3 What are the main symptoms?

The main symptoms are hip pain and a limp. Hip pain may be present as pain in the groin, upper thigh or occasionally in the knee, usually of sudden onset. The most common manifestation is a child awakening with a limp or refusing to walk.

7.4 How is it diagnosed?

The physical examination is characteristic: a limp with decreased and painful hip motion in an afebrile child older than 3 years of age, who otherwise does not appear ill. Both hips are affected in 5% of cases. Hip x-rays generally have normal findings and are not usually required. In contrast, hip ultrasound is very useful in detecting hip synovitis.

7.5 How can we treat it?

The basis of the treatment is rest, which should be proportionate to the degree of pain. Non-steroidal anti-inflammatory drugs can help to decrease pain and inflammation. The condition usually resolves after an average of 6-8 days.

7.6 What is the prognosis?

Prognosis is excellent with full recovery in 100% of children (it is transient by definition). If symptoms persist for more than 10 days, a different disease should be considered. It is not uncommon to develop new episodes of transient synovitis; these episodes are usually milder

and shorter than the first one.

8. Patellofemoral pain - knee pain

8.1 What is it?

Patellofemoral pain is the most common paediatric Overuse Syndrome. Disorders of this group result from repetitive motion or sustained exercise-related injury to a particular part of the body, in particular the joints and tendons. These disorders are much more common in adults (tennis or golf elbow, carpal tunnel syndrome, etc.) than in children. Patellofemoral pain refers to the development of anterior knee pain with activities that place additional load over the patellofemoral joint (the joint formed by the knee cap (patella) and the lower part of the thigh bone or femur).
When the knee pain is accompanied by changes in the inner surface tissue (cartilage) of the patella, the medical term "chondromalacia of the patella" or "chondromalacia patellae" is used.
There are many synonyms for patellofemoral pain: patellofemoral syndrome, anterior knee pain, chondromalacia of patella, chondromalacia patellae.

8.2 How common is it?

It is very uncommon in children up to 8 years, becoming progressively more common in adolescents. Patellofemoral pain is more common in girls. It can also be more common in children with significant angulation of the knees such as knock-knees (genu valgum) or bow legs (genu varum), as well as in those with diseases of the patella due to misalignment and instability.

8.3 What are the main symptoms?

The characteristic symptoms are anterior knee pain that worsens with activities such as running, climbing up or down stairs, squatting or jumping. Pain is also worsened by a prolonged sitting position with the knee bent.

8.4 How is it diagnosed?

Patellofemoral pain in healthy children is a clinical diagnosis (lab test or imaging studies are not necessary). Pain may be reproduced by compression of the knee cap or by restraining the upward movement of the patella when the thigh muscle (quadriceps) is contracted.

8.5 How can we treat it?

In most children with no associated diseases (such as angulation disorders of the knees or patellar instability), patellofemoral pain is a benign condition that resolves by itself. If the pain interferes with sports or with daily activities, initiation of a programme of quadriceps strengthening may be of help. Application of cold packs may relieve pain after exercise.

8.6 What about everyday life?

Children should lead a normal life. Their level of physical activity should be adjusted to keep them pain-free. Very active children may use a knee sleeve with a patellar strap.

9. Slipped Capital Femoral Epiphysis

9.1 What is it?

This condition is a displacement of the femoral head through the growth plate; the cause is unknown. The growth plate is a slice of cartilage sandwiched between bone tissues in the femoral head. It is the weakest part of the bones and allows them to grow. Once the plate is mineralized and becomes bone itself, the bones stop growing.

9.2 How common is it?

It is an uncommon disease that affects 3-10 in 100,000 children. It is more frequent in adolescents and in boys. Obesity seems to be a predisposing factor.

9.3 What are the main symptoms?

Limp and hip pain with decreased mobility of the hip are the main symptoms. Pain may be felt in the upper (two thirds) or lower (one third) thigh, and it increases with physical activity. In 15% of children, the disease affects both hips.

9.4 How is it diagnosed?

The physical examination is characteristic, with decreased hip mobility. The diagnosis is confirmed by X-rays, preferably in the axial ("frog-leg") view.

9.5 How can we treat it?

This condition is considered an orthopaedic emergency and requires surgical pinning (stabilization of the femoral head by insertion of pins to keep it in place).

9.6 What is the prognosis?

It depends on how long the femoral head has been in the slipped position before the diagnosis and on the degree of slippage. It varies from child to child.

10. Osteochondrosis (Synonyms: osteonecrosis, avascular necrosis)

10.1 What is it?

The word "osteochondrosis" means "bone death". It refers to a diverse group of diseases of unknown cause, characterised by interruption of blood flow to the ossification centre of the affected bones. At birth, bones are mostly made of cartilage, a softer tissue that is replaced over time by a more mineralized and resistant tissue (the bone). This replacement begins at specific sites within each bone, areas known as ossification centres, spreading out to the rest of the bone over time. Pain is the main symptom of these disorders. Depending on the bone affected, the disease receives different names.
The diagnosis is confirmed by imaging studies. X-rays show, in sequence, fragmentation ("islands" within the bone), collapse

(breakdown), sclerosis (increased density, the bone looks "whiter" on the films) and, frequently, re-ossification (new bone formation) with reconstitution of the bone contour.
Although it may sound like a serious disease, it is quite common in children and, with the possible exception of extensive involvement of the hip, it has an excellent prognosis. Some forms of osteochondrosis are so frequent that they are considered a normal variation of bone development (Sever´s disease). Others may be included in the group of "overuse syndromes" (Osgood-Schlatter, Sinding-Larsen-Johansson diseases).

10.2 Legg-Calvé-Perthes Disease

10.2.1 What is it?

This disease involves avascular necrosis of the femoral head (the part of the thigh bone closest to the hip).

10.2.2 How common is it?

It is not a common disease, reported in 1/10,000 children. It is more frequent in boys (4/5 boys for every 1 girl) between the ages of 3 and 12 years and occurs particularly in children 4 to 9 years old.

10.2.3 What are the main symptoms?

Most children present with a limp and variable degrees of hip pain. Sometimes pain may not be present at all. Only one hip is usually involved but in about 10% of cases the disease is bilateral.

10.2.4 How is it diagnosed?

The mobility of the hip is impaired and may be painful. X-rays may yield normal findings at the beginning but later show the progression described in the introduction. Bone scans and magnetic resonance imaging detect the disease earlier than X-rays.

10.2.5 How can we treat it?

Children with Legg-Calvé-Perthes disease should always be referred to a paediatric orthopaedic department. Imaging is essential for diagnosis. Treatment depends on the severity of the disease. In very mild cases, observation may be sufficient, as the bone heals by itself with little damage.
In more severe cases, the goal of therapy is to contain the affected femoral head within the hip joint, so that when the formation of new bone starts, the femoral head recovers its spherical form.
This goal may be achieved to a variable degree by wearing an abduction brace (younger children) or by surgically reshaping the femur (osteotomy, cutting a wedge of bone to keep the femoral head in a better position) (in older children).

10.2.6 What is the prognosis?

The prognosis depends on the extent of femoral head involvement (the less, the better) as well as on the age of the child (better if under 6). Full recovery takes 2 to 4 years. Overall, about two-thirds of affected hips have a good long-term anatomical and functional outcome.

10.2.7 What about everyday life?

The limitations for daily activities depend on the treatment applied. Children under observation should avoid heavy impact to the hip (jumping, running). However, they should carry on with an otherwise normal school life and participate in all other activities that do not involve heavy weight bearing.

10.3 Osgood-Schlatter Disease

This condition results from repeated trauma to the ossification centre of the tibial tuberosity (a small bone crest present in the upper leg) by the patellar tendon. It is present in about 1% of adolescents and is more common in individuals who play sports.
Pain worsens with activities such as running, jumping, going up or down stairs and kneeling. The diagnosis is established by the physical examination, with a very characteristic tenderness or pain, sometimes accompanied by swelling, at the insertion of the patellar tendon into the tibia.

X-rays might be normal or show small fragments of bone in the tibial tuberosity. Treatment involves adjusting the level of activity to keep patients pain-free, applying cold packs after sports and rest. The condition resolves over time.

10.4 Sever´s Disease

This condition is also called "calcaneal apophysitis". It is an osteochondrosis of the calcaneal apophysis of the heel bone, probably related to the traction of Achilles tendon.
It is one of the most common causes of heel pain in children and adolescents. Like other forms of osteochondrosis, Sever´s disease is activity-related and more common in males. Its onset is usually around 7-10 years of age, with heel pain and occasionally a limp after exercise. The diagnosis is made by the clinical examination. There is no need for therapy other than to adjust the level of activity to keep the child pain-free and, if this approach does not work, to use a heel cushion. The condition resolves over time.

10.5 Freiberg´s Disease

This condition describes the osteonecrosis of the head of the second metatarsal bone in the foot. Its cause is probably trauma. It is uncommon and most cases involve adolescent girls. The pain increases with physical activity. The physical examination shows tenderness under the second metatarsal head and occasionally swelling. The diagnosis is confirmed by X-rays, although it may take two weeks from the beginning of the symptoms before changes are detectable. Treatment includes rest and a metatarsal pad.

10.6 Scheuermann´s Disease

Scheuermann’s disease or "juvenile kyphosis (roundback)" is an osteonecrosis of the ring apophysis of the vertebral body (the bone on the periphery of the top and bottom of each vertebra). It is more common in adolescent boys. Most children with this condition have poor posture, with or without back pain. The pain is activity-related and may be relieved by rest.
The diagnosis is suspected upon examination (sharp angulation in the

back) and confirmed by X-rays.
To receive a diagnosis of Scheuermann´s disease, the child must have irregularities of the vertebral plates and anterior "wedging" of 5 degrees in at least three consecutive vertebrae.
Scheuermann´s disease usually does not require therapy other than to adjust the level of activity of the child, observation and, in severe cases, bracing.