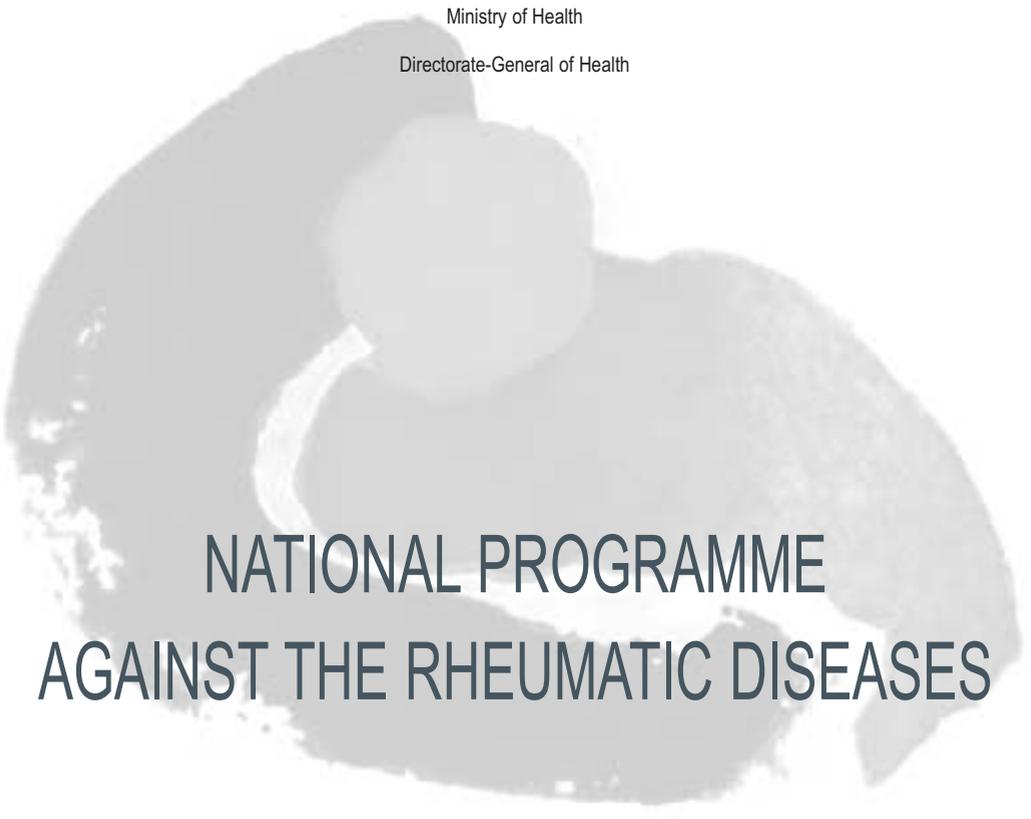


# NATIONAL PROGRAMME AGAINST RHEUMATIC DISEASES



DIRECTORATE-GENERAL OF HEALTH

Ministry of Health  
Directorate-General of Health



# NATIONAL PROGRAMME AGAINST THE RHEUMATIC DISEASES

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## ABBREVIATIONS

GCA:	Giant cell arteritis
IJA:	Idiopathic juvenile arthritis
NSAID:	Nonsteroidal anti-inflammatory drugs
PA:	Psoriatic arthritis
RA:	Rheumatoid arthritis
ReA:	Reactive arthritis
BASDAI:	Bath ankylosing spondylitis disease activity index
BASFI:	Bath ankylosing spondylitis functional index
BASG:	Bath ankylosing spondylitis global score
BASMI:	Bath ankylosing spondylitis metrology index
CK:	Creatine kinase
PHC:	Primary health care
DEXA:	Dual-energy X-ray absorptiometry
RD:	Rheumatic disease(s)
PRD:	Periarticular rheumatic diseases
SRD:	Systemic rheumatic diseases
AS:	Ankylosing spondylitis
CDE:	Complementary diagnostic exams
SNSA:	Seronegative spondylarthropathy
SAIBD:	Spondylitis associated to inflammatory bowel disease
uSpA:	Undifferentiated spondylarthropathy
EMG:	Electromyogram
SSc:	Systemic sclerosis
FM:	Fibromyalgia
PFF:	Proximal femur fractures
UG:	Uric gout
WG:	Wegener's granulomatosis
M:	Man
HAQ-s:	Health assessment questionnaire – short form

SLE:	Systemic lupus erythematosus
WRMI:	Work related musculoskeletal disorders
W:	Woman
M-S:	Musculoskeletal
MTX:	Methotrexate
OA:	Osteoarthritis
WHO:	World Health Organization
OP:	Osteoporosis
PAN:	Polyarteritis nodosa
CRP:	C-reactive protein
PM/DM:	Polymyositis / dermatomyositis
POA:	Polio-osteoarthritis
NMR:	Nuclear magnetic resonance
APAS:	Antiphospholipid antibodies syndrome
SAARD:	Slow-acting antirheumatic drugs
SAPHO:	Synovitis, acne, pustulosis, hyperostosis and osteitis
CNS:	Central nervous system
SS:	Sjögren's syndrome
RS:	Reiter's syndrome
CAT:	ComputerisedComputerized axial tomography
TNFa:	Tumour Tumor Necrosis factor alpha
TSH:	Thyroid stimulating hormone

## INTRODUCTION

The rheumatic diseases, aside from being the most common group of diseases of the human race in developed countries, must also be viewed as an important social and economic problem whose negative impact on public health has a tendency to increase considering current lifestyles and longer life expectancy.

This National Programme Against the Rheumatic Diseases, presented here, is an integral part of the National Health Plan for 2004 – 2010 and should also be considered a contribution of the Ministry of Health to the international “Bone and Joint Decade 2000 – 2010” movement, an initiative of the United Nations, supported by the World Health Organization. In accordance with the National Health Plan, this programme aims to invert the growing tendency of this problem and calls for a comprehensive and articulate approach of the health services aimed at reducing, among the Portuguese population, the risk of contracting these diseases and to provide suitable treatment and rehabilitation.

This programme’s investment not only in primary preventive measures but essentially, in secondary and tertiary prevention makes a special appeal to all health provider services to join forces in order to quickly obtain clear health gains in rheumatology

The National Programme Against the Rheumatic Diseases will be applied essentially by the implementation of strategies of intervention, education, data collection and analyses. Support instruments will be developed, replicated and creatively adapted to the various towns and regions as required by the specific characteristics of each location.

To meet these strategies, the Directorate-General of Health selected the Portuguese Society of Rheumatology as its permanent scientific liaison. Nevertheless, when necessary it may also collaborate with the Portuguese Orthopaedics and Traumatology Society, the Portuguese Physical and Rehabilitation Medicine Society, the Portuguese General Physicians Association, the Portuguese Work Medicine Association, with other scientific associations, associations of patients and of professional orders.

It is common for one person to have multiple chronic pathologies that interact and are mutually inductive. It is therefore essential that the recently created National Programme Against the Rheumatic Diseases carry out measures within a perspective of collaboration and communication with the current health care programmes and services, in particular primary care, hospital care and ongoing care, and with the national health programmes in effect.

## CONTEXT

The rheumatic diseases may be defined as *diseases and functional disorders of the musculoskeletal system caused by a non-traumatic source*. Over one hundred rheumatic entities exist, with various subtypes, which include inflammatory diseases of the musculoskeletal system, of the “connective tissues” and of the vascular system, degenerative diseases of peripheral joints and of the spine, metabolic diseases of the bone and joints, disorders of soft periarticular tissue and diseases of other organs and/or systems related with the previous ones.

These diseases may be acute, recurrent or chronic, and affect persons of all ages. They are frequently the cause of disability and of notable asymmetries in the access to benefits granted under a special regime. Rheumatic diseases, when not diagnosed or when not subject to correct and timely treatment may cause serious and unnecessary physical, psychological, family, social and economic repercussions.

The clinical symptoms of rheumatic diseases such as pain, swelling and limited motion are very common in the general population. The prevalence of these symptoms is higher in women and in older age groups as well as in those with lower income and education. These symptoms are most frequently caused by osteoarthritis, back pain, periarticular disorders, including work-related musculoskeletal lesions, osteoporosis, fibromyalgia, microcrystalline arthropathies, rheumatoid arthritis, spondyloarthropathies, systemic rheumatic diseases and idiopathic juvenile arthritis.

Overall, the rheumatic diseases are one of the main causes of both direct health care costs – consultations, medicines or rehabilitation care – as well as indirect costs.

The true dimension of the problem, caused by the rheumatic diseases in Portugal is not rigorously known. Although mortality caused by these diseases is low, at least 30% of the population refer having musculoskeletal symptoms, whereby: 20% have a significant problem, that is, they are sick; 7% must limit day-to-day activities, that is, have a disability; and 0.5% depend on a third party, which means they are handicap;

Epidemiological studies, conducted since 1976 in Portugal reveal similar results, which gives them scientific consistency, confidence and value. From the results it may be inferred that the rheumatic diseases in Portugal have a prevalence of about 20% to 30%; are responsible for 16% to 23% of general practice consultations; are the 2<sup>nd</sup> or 3<sup>rd</sup> reason for which patients take medicines; are the 1<sup>st</sup> cause of temporary disability; are responsible for 17% of definitive bedridden patients, 26% of persons requiring wheelchairs, 30% of persons restricted to mobility at home; 40% to 60% of situations of prolonged disability for certain day-to-day activities; are responsible for 43% of work absenteeism due to illness and are the greatest cause of early retirement due to illness, that is, 35% to 41% of the total.

Therefore, it must be kept in mind that:

- a) Osteoarthritis is the main cause of disability in the elderly. In this age group, it is frequently associated with other diseases that are often disabling, and affects essential joints such as those of the hands, knees, hips, back and feet.
- b) Back pain, or pain in the spine, whether of a degenerative, infectious, inflammatory, metabolic or neoplastic nature, are the most common complaints caused by rheumatism and are one of the main reasons for disability before age 45.
- c) Periarticular disorders, which are frequently caused by lesions arising from repeated work-related trauma, represent over half of occupational diseases.
- d) Osteoporosis, which reduces bone resistance, increases the likelihood of fractures caused by low-intensity trauma, causes, in Portugal, 40,000 fractures every year, of which 8,500 are of the proximal femur, and are estimated to be responsible for over 50 million euros merely in hospital care costs. These fractures are one of the main causes of morbidity and mortality.
- e) Fibromyalgia, a non-inflammatory chronic musculoskeletal syndrome of unknown cause, gives rise to physical and emotional disability, sometimes serious, due to the general pain, fatigue, quantitative and qualitative sleep and cognitive disorders.
- f) Microcrystalline arthropathy, particularly uric gout, is a frequent cause of renal dysfunction.
- g) Rheumatoid arthritis causes severe morbidity and disability in young age groups, reducing life expectancy, in more serious forms, by about 10 years.

- h) Spondyloarthropathies, including ankylosing spondylitis, psoriatic arthritis, reactive arthritis and the arthropathy associated with chronic inflammatory bowel diseases will mainly affect the spine and sacroiliac joints, causing major disabilities in younger age groups.
- i) Systemic rheumatic diseases, comprising a heterogeneous group of diseases and syndromes, such as systemic lupus erythematosus, Sjögren's syndrome, systemic sclerosis, polymyositis / dermatomyositis and the wide number of vasculitis affect young adults and may have a serious prognosis.
- j) Juvenile idiopathic arthritis are of the most common chronic diseases affecting children and adolescents and are a major cause of disability, ocular disease and of failure rates at schools because of absenteeism.

The Rheumatic diseases thus comprise a wide nosological group that, overall, causes high morbidity, high temporary disability, frequent absenteeism from work, early irreversible disability resulting in greater early retirement due to invalidity, lower life expectancy and negative economic and social impacts. However, in Portugal rheumatology is the clinical specialty with the lowest hospital expression, since in the mainland and autonomous regions, there are only eight rheumatology services or units in public hospitals, in addition to a rheumatology service in a military hospital and the Portuguese Rheumatology Institute, a non-profit private institute dedicated exclusively to rendering rheumatoid care. In fact, Portugal's National Health Care Service has only 75 rheumatologists and 26 interneees under training, which clearly places us way behind international standards in this matter.

It has been proven that a correct early diagnosis followed by appropriate and timely treatment of rheumatic diseases significantly reduces their effects on physical disability and the need for treatment. It is therefore necessary to increase awareness and education about these diseases among health care workers of various levels.

Moreover, until now and contrary to other chronic incapacitating diseases, the country does not have any national plan or programme to promote prevention in the rheumatic diseases at the various levels. This situation must be changed by launching this National Programme Against Rheumatic Diseases to ensure that persons suffering from rheumatism will have their diseases diagnosed and treated appropriately. The programme's measures will improve the quality of life of the respective patients and also entail substantial financial and resource savings for them and for society.

## GOALS

The National Programme Against Rheumatic Diseases aims to meet the following goals:

### GENERAL

1. Control morbidity and mortality caused by rheumatic diseases.
2. Improve the quality of life of rheumatic patients.
3. Cut down on costs associated to rheumatic diseases.

### SPECIFIC

1. Determine the prevalence of rheumatic diseases covered by this programme.
2. Determine the incidence of periarticular and work-related musculoskeletal disorders.
3. Determine the incidence of low back pain.
4. Determine the incidence of osteoporotic fractures.
5. Determine criteria for identifying each rheumatic patient's needs for benefits under a special regime and to propose measures to rationalize the assignment of those benefits.

## TIME FRAME

The National Programme Against the Rheumatic Diseases aims to cover a 10-year period after having been approved, without loss to any corrections deemed appropriate for its progress.

The programme will cover two stages:

- a) Implementation stage, covering the first five years after its approval.
- b) Consolidation stage, from the sixth year on.

## TARGET POPULATION

In addition to the general population, as a priority and *ab initio*, this programme will be developed to meet the needs of the following target populations that are particularly at risk:

1. Children and adolescents.
2. Post-menopausal women.
3. The elderly.
4. Workers with repetitive physical activities or with ergonomic disorders.
5. Those practicing amateur or occasional sports.

## STRATEGIES

The National Programme Against the Rheumatic Diseases must be developed by implementing the following strategies at a national, regional and local level:

### INTERVENTION STRATEGIES

- E1 Creation and development of rheumatology hospital services and/or units.
- E2 Production and disclosure of technical guidelines about the diagnosis, follow-up and referral of rheumatic patients, particularly for:
  - a) Osteoarthritis
  - b) Back pain
  - c) Periarticular disorders
  - d) Work-related musculoskeletal disorders
  - e) Osteoporosis
  - f) Fibromyalgia
  - g) Microcrystalline arthropathies
  - h) Rheumatoid arthritis

- i) Spondyloarthropathies
  - j) Systemic rheumatic diseases
  - k) Juvenile idiopathic arthritis
- E3 School health teams will develop and disseminate technical guidelines for identifying children with modifiable risk factors that may evolve to musculoskeletal diseases, refer them in a timely manner to specialized rheumatology units, and integrate these units in the school environment.
- E4 School health teams will develop and disseminate technical guidelines about the ergonomics in the school environment.
- E5 Development and dissemination of technical guidelines for timely transversal screening of disorders of the musculoskeletal stability and dynamics of 6-year-old children.
- E6 Periodic disclosure to health care workers about where general rheumatology and children's rheumatology consultations are available.
- E7 Production and disclosure, by occupational health services, of technical guidelines about work environment ergonomics.
- E8 Production and disclosure, by Health Centres and the Ministry of Social Security and Labour, of technical guidelines about the prevention of falls in the elderly.
- E9 Development of a technical proposal for dietary supplementation with vitamin D and calcium in the elderly.
- E10 Validation of criteria for evaluating the functionality of rheumatic patients.
- E11 Development of a proposal for a model of stratifying rheumatic patients to access benefits granted under a special regime.

## TRAINING STRATEGIES

- E12 Promote medical schools to increase the number of pre-and post-graduate training hours in rheumatology.
- E13 Promote the National Commission of Medical Internships and hospital management boards to increase the number of positions for the Complementary Internship in Rheumatology.

- E14 Promote compulsory rheumatology training in the Complementary Internship of General and Family Practice.
- E15 Promote specific training, on the musculoskeletal system and the rheumatic diseases, to non-physician health care providers, senior sports technicians and teachers of various education levels.
- E16 Development of pedagogic instruments, targeted towards health care providers, for early detection of inflammatory arthropathy and systemic rheumatic diseases.
- E17 Raise the awareness of entrepreneurs and other employers, as well as of labour unions and other labour associations about the need to prevent periarticular rheumatic diseases and work-related musculoskeletal injuries and the adoption of measures that increase the adequacy of labour activities to each patient's limitations.
- E18 Raise the awareness of health providers about the advantages of assigning benefits under a special regime based on the specific needs of each rheumatic patient.
- E19 Develop multisectorial partnerships for disclosing general information, to the public at large, about rheumatic diseases and their prevention and, in particular, about:
  - a) Osteoarthritis and back pain
  - b) Fibromyalgia
  - c) Hyperuricemia and uric gout
  - d) Inflammatory arthropathies and periarticular rheumatic diseases
  - e) Good bone health habits

## STRATEGIES FOR COLLECTING AND ANALYSING INFORMATION

Development of multisectorial partnerships to create an observatory for the rheumatic diseases that:

- E20 Include systems for data collection that permit the collection and analyses of data on prevalence and incidence of the rheumatic diseases, as well as on temporary and definitive capacity and work absenteeism caused by these diseases or by their complications.
- E21 Monitor health gains resulting from the actions of this programme.

## CALENDAR

Implementation	1 <sup>st</sup> year				2 <sup>nd</sup> year				3 <sup>rd</sup> year				4 <sup>th</sup> year				5 <sup>th</sup> year			
Stage	Trimestrial				Trimestrial				Trimestrial				Trimestrial				Trimestrial			
Strategies	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20
E1	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E2	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E3	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E4	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E5	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E6	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E8	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E9	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E10	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E11	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E12	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E13	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E14	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E15	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E16	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E17	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E18	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E19	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E20	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	
E21	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	█	

## FOLLOW-UP AND EVALUATION

Annual Monitoring and evaluation of the National Programme Against the Rheumatic Diseases, implemented by the health care services, will be accomplished by the departments supervised by the Regional Health Services

This programme is nationally coordinated by the Directorate-General of Health through a National Coordination Commission to be formed by a Government Order.

The National Programme Against the Rheumatic Diseases will be monitored to determine whether it has attained health gains based on the following indicators, according to gender:

### PREVALENCE

- a) Osteoarthritis
- b) Back pain
- c) Periarticular rheumatic diseases
- d) Arthropathy and uric nephropathy
- e) Blindness associated to juvenile idiopathic arthritis
- f) Work-related musculoskeletal injuries, per occupation area

### INCIDENCE

- a) Low back pain
- b) Osteoporotic vertebral fractures and hip fractures
- c) Periarticular rheumatic diseases
- d) Work-related musculoskeletal injuries per occupational area

## DISABILITY

- a) Number of days of temporary disability caused by a rheumatic disease
- b) Number of days of work absenteeism caused by a rheumatic disease
- c) Median age of definitive disability caused by a rheumatic disease

## MORTALITY

- a) Caused by inflammatory arthropathy
- b) Caused by systemic rheumatic diseases

## TECHNICAL GUIDELINES

### OSTEOARTHRITIS

#### DEFINITION

Osteoarthritis (OA), or simply arthritis, may be defined as a joint disease resulting from the failure of various repair mechanisms to multiple insults and injuries suffered by the joint. From an anatomical and pathological perspective, there is focal destruction of cartilage and a reaction by the subchondral bone, but the process involves the entire joint, including the capsule, synovial membrane, ligaments and periarticular muscles. Clinically, it presents with joint pain, stiffness and limitation of motion, and characteristically there is no systemic repercussion.

The most commonly affected joints in OA are:

- a) cervical and lumbar segments of the spine;
- b) first carpometacarpal joint of the hand;
- c) proximal and distal interphalangeal joints of the hand;
- d) hips;

- e) knees;
- f) first metatarsophalangeal joint.

OA is usually classified as primary or idiopathic and secondary. Most cases are classified as idiopathic, while secondary forms of OA may be grouped into:

- a) metabolic and endocrine, that include diseases caused by the deposition of calcium crystals, ochronosis, acromegaly and hemochromatosis;
- b) anatomical, that may include hypermobility syndromes, axial shifting of the limbs, serious scolioses, epiphyseal dysplasia, Perthes disease, congenital hip dislocation;
- c) traumatic, ranging from acute joint trauma to chronic trauma, occupational or sports-related, surgery such as meniscectomy.

According to the pattern of joint involvement, OA may be mono, oligo or polyarticular. The term generalized OA or poliosteoarthritis is applied when three or more regions are involved by the disease. Certain clinical patterns also justify specific nomenclature such as erosive OA of the hands and rapidly destructive chondrolysis that can affect the knee or hip.

## RISK FACTORS

Risk factors may be divided into non-modifiable and potentially modifiable. The former include age, in which there is a very clear link between OA and ageing, although, from an anatomical and pathological perspective, the disease is clearly distinguished from the joint's senescence process, race in certain OA locations, gender being more common in women, metabolic or endocrine diseases and inflammatory arthropathies.

Of greater importance are modifiable risk factors such as obesity, major trauma on joints and joint overload resulting from occupational or leisure activities, anatomical defects, the reduction of quadriceps force (in knee OA) and proprioceptive defects. Genetic factors, important in hand and finger OA and in the forms of POA, may soon be included in this list of modifiable factors.

The higher risk population is made up mostly of the elderly, and women, the obese, those whose joints are subject to overloads due to work or sports,

those with anatomical defects that affect the normal biomechanics of joints and those that suffer from other joint and bone diseases, including trauma.

### PREVENTION

OA evolution is slow measured in years or even decades. The disease may become stable, be slowly progressive, with or without flares of acute symptoms and even inflammatory signs that normally coincide with a radiographic deterioration. Nevertheless, there are rapidly developing forms, in the hip or knee, which are called rapidly destructive chondrolysis that destroys nearly all of the cartilage in a few months and must be actively treated.

The prognosis of OA varies greatly, depending on the affected joint. In the hands, thumb base OA (carpometacarpal joint OA) may be very debilitating, while interphalangeal joint OA may cause an important deformity and pain, in flares, in the first years, but normally does not compromise hand function. In the spine, cervical OA may also be very symptomatic, especially in the beginning, or cause neurological complications, but more commonly it stabilises and even improves in terms of symptoms. In contrast, OA of the lumbar spine is almost always debilitating, with major occupational and socioeconomic repercussions, and may also include neurological complications that are difficult to treat. Knee and hip OA may also evolve very negatively in terms of symptoms and function, and clearly limits patient mobility. However, the availability of total prosthesis for these joints have greatly improved the prognosis.

It is almost always inevitable that symptoms – pain, stiffness – amplify limited motion caused by the anatomical changes in the joint tissues, whereby the patient goes through a voluntary process of autolimiting joint function, which decreases pain but reduces quality of life.

OA is diagnosed by typical radiographic findings in patients complaining of joint pain, generally mechanical in nature, stiffness and, almost always, to a greater or lesser extent, limitation of joint motion and function. These radiographic findings, which are the basis for diagnosis, consist of joint space narrowing reflecting cartilage thinning, bony subchondral sclerosis and growth of marginal osteophytes. When these findings are present, the arthritic process is already advanced and thus conventional x-ray diagnosis is never made early enough.

Early disease detection could be achieved by arthroscopy, under the limitations of an invasive diagnostic method, or by magnetic resonance, which is costly and of difficult access. Additional difficulties are posed by a common dissociation between the intensity of clinical manifestations and the severity of x-ray findings. It is not rare for typical OA signs to be found in x-rays when there are no symptoms and the disease's natural history is, often, and especially in the beginning, unpredictable.

The primary prevention of OA must be based on correcting identifiable risk factors:

- a) Treating obesity;
- b) correcting joint abnormalities, congenital or developmental, axial deviations and leg length discrepancy;
- c) avoiding joint overload and repetitive trauma, even at small ranges, particularly in jobs requiring flexion of the trunk and lifting weights;
- d) quadriceps strengthening;
- e) attempt to correct proprioceptive defects that determine repetitive biomechanical defects during lifetime.

Secondary prevention is very limited by the already mentioned difficulties in early diagnosis.

Tertiary prevention requires the use of pharmacological modalities that modify the disease's natural history and the development of other drugs aimed at delaying disease course. Surgical treatment is very important, especially placing hip and knee prosthesis in advanced cases.

## TREATMENT

OA treatment goals are as follows:

- a) Patient education;
- b) Treat symptoms;
- c) Minimise disability;
- d) Slow disease progression.

Patient education and information is essential for obtaining cooperation, and various scientific studies have shown its effectiveness in improving symptoms and quality of life. Implementation of measures to help obese patients to lose weight may be included here.

Rehabilitation includes a series of techniques that range from the application of physical agents, for symptomatic relief, hydrotherapy, thermal modalities, support devices (for activities of daily living, walking aids and other orthotics), teaching joint protection techniques and energy saving methods. Kinesiotherapy, on its own or combined with hydrotherapy, is effective in relieving symptoms and may delay disease progression.

Pharmacologic modalities to treat symptoms may be topical, systemic, intra-articular or periarticular. The latter is very useful for relieving frequent inflammatory conditions of the soft tissues surrounding joints. There are also drugs for slow and lasting relief of symptoms and that may slow down disease progression.

Joint lavage has also been proven to be effective in improving symptoms of OA.

Lastly, in a somewhat early stage, orthopaedic surgery may correct anatomical defects or joint deformities that imply overload and, in a later stage, partial or total joint replacement with prosthesis.

OA, a multi-faceted disease in etiology, clinical presentation and evolution, requires an individualised and diversified therapeutic programme through a multidisciplinary team that includes a general practitioner, rheumatologist, physiatrist, orthopaedic surgeon, nurse and therapists. This concept must also include social support, psychological counselling, occupational medicine and professional reclassification.

#### FOLLOW-UP

Follow-up of OA treatment is essentially clinical, including the evaluation of symptoms and joint function, since in advanced stages of the disease, where pain is a major factor, there is a natural tendency to voluntarily restrict motion and, as such, to reduce symptoms, which misleads the assessor. On the other hand, radiology is not very sensitive, so, besides its use in diagnosis, x-rays are only necessary when there is a modification of symptoms or of the physical exam.

Follow-up must include:

- a) overall evaluation of the disease by the patient and the physician;
- b) a quantitative evaluation of pain and function;
- c) a count of inflammatory flares;
- d) a physical exam with special emphasis on joint evaluation.

Follow-up should be performed every six months, or after longer intervals if the situation remains stable or shorter if changes arise.

#### PATIENT REFERRAL

Most patients with OA may be diagnosed, treated and monitored in primary health care.

The following patients must be referred to rheumatologists:

- a) patients whose diagnosis is questionable, for example, when there are symptoms suggesting the disease without clear radiographic changes;
- b) when there are doubts about the possible etiology;
- c) when there is an important, persistent or repeated joint effusion;
- d) when the disease evolves rapidly (clinically or radiographically);
- e) whenever the response to the therapy is considered insufficient.

The following patients must be referred to an orthopaedic surgeon:

- a) patients requiring anatomical repair to improve joint biomechanics;
- b) patients whose disease has evolved to a stage at which a joint prosthesis may be necessary.

The following patients must be referred to a physiatrist:

- a) patients who may benefit from the application of physical agents for symptomatic relief;
- b) patients needing a programme of kinesiotherapy for muscle strengthening and/or to improve the biomechanics of the joint.

## BACK PAIN

### DEFINITION

Back or spine pain is a very common symptom, in which the cervical and lumbar segments are the most frequently affected since these have greater mobility.

In most cases, neck pain is caused by degenerative deterioration or a functional change of musculoligamental structures. Radiographic changes of cervical spine OA are very common, affecting more than half of persons over age 40, but are asymptomatic in most cases.

Low back pain is a major public health problem, particularly in industrialised countries where it is common and affects part of the active population, leading to work absenteeism and a significant loss of productivity. The socioeconomic impact of disability due to low back pain has increased exponentially in these countries, and the most significant costs are incurred by its chronic forms.

Low back pain is a symptom and not a disease. It has multiple causes that are not fully known: the same symptom, pain, may result from situations of distinct physiopathology. It is labelled according to duration, to whether it is acute or chronic, in which case those lasting more than 3 months are chronic. The chronic form represents only 7% of low back pain, but it is estimated to be responsible for over 75% of costs arising from this ailment.

According to its etiopathogenic mechanism, low back pain may be classified as mechanical or non-mechanical. Mechanical low or common back pain is defined as pain caused by overload or overuse of a normal anatomic structure, secondary to trauma or to deformity, and may have an underlying degenerative disk disease or an alteration of the posterior interapophyseal joints. The non-mechanical, atypical or symptomatic forms may result from heterogeneous situations – inflammatory, infectious, metabolic or cancerous – or referred pain from an extra-vertebral origin. Traumatic and psychogenic causes must also be taken into account.

## RISK FACTORS

The main risk factors include:

- a) age over 45;
- b) female;
- c) smoking;
- d) alcoholism;
- e) occupational factors;
- f) psychological factors.

The early detection of the following factors is very important for predicting whether the problem will become chronic:

- a) age;
- b) duration of the initial episode;
- c) duration of absence of work;
- d) relapses;
- e) hospitalisation;
- f) low educational level;
- g) low income;
- h) poor family environment;
- i) prior depression and anxiety;
- j) poor working conditions;
- k) poor job qualifications and dissatisfaction;
- l) precarious work;
- m) a conflict resulting from a work accident;
- n) unsuitable use of diagnostic and therapeutic resources.

## PREVENTION

Low back pain is a common symptom affecting the general population, and it is estimated that 60% to 80% of people are affected by a bout of low back pain during their life. The risk of low back pain increases in certain jobs, particularly those requiring strenuous physical activity or prolonged postures in which the vertebral column is bending and/or rotating. It is a major occupational problem, relapses are frequent and result, in most cases, from overload or poor use of the vertebral and paravertebral structures.

In the approach to low back pain, the first goal is always to distinguish between the mechanical and non-mechanical causes of pain. That is, one must identify situations of infection, inflammatory disease, tumour, osteoporotic fracture or an extra-vertebral pathology needing specific treatment.

The initial approach to low back pain is based essentially on clinical information. A careful history and physical exam are generally sufficient to diagnose common low back pain and/or suspicion of other origins.

Although degenerative disorders are very prevalent, the patient's complaints rarely have a well identified cause, that is, no good anatomical-clinical correlation exists.

The patient's socio-professional and psychological context must be taken into account in the initial approach to determine risk factors in the development to a chronic state.

Primary, secondary or tertiary prevention is meant for the general public, with special focus on some professional groups. Particular attention must be given to the prevention of chronic conditions and recurrences.

Primary prevention, which aims to avoid low back pain, is of particular interest in:

- a) schools through postural and gestural education, since poor use of the back begins early in life;
- b) sports played during the growth stage;
- c) the workplace.

The goal of secondary prevention is to reduce the severity and relapse rate of painful episodes. Its application is important in schools where educational measures must be implemented to teach the essential rules of vertebral well-being by avoiding useless overloads. Education should be applied to small groups by a multidisciplinary team and includes elementary theoretical notions on the anatomy, physiology and pathology of the back, exercise and posture and, in some cases, behavioural therapy.

Tertiary prevention is of interest in chronic low back pain, even in individuals who are most disabled. It should aim to restore social and occupational activities, improve muscular capacity and physical shape in general. Tertiary prevention is based on training to reduce disability's duration and for preventing a chronic state.

## TREATMENT

No single form of therapy is effective for all forms of low back pain. The guidelines to be followed depend on whether the situation is acute or chronic, on the presence or absence of radicular pain, on the origin of the pain (discal, posterior interapophyseal or musculotendonous) and on the patient's socio-professional and psychological context.

In acute situations, excluding cases of surgical emergencies, the treatment aims to alleviate pain and may include:

- a) short course of rest;
- b) analgesics;
- c) NSAIDs;
- d) Muscle relaxants in the case of muscular contraction.

A fast and progressive resumption of normal activities is essential, whereas continued rest is not only useless but also delays recovery. Local therapy, such as epidural injections, may be indicated for rare cases of radicular pain, but only after conservative treatment has failed after 4 to 8 weeks; they are of no interest for low back pain without sciatic pain.

To prevent relapses and recurrences, dorsolumbar support bands may be used in some activities, abdominal and vertebral strengthening exercises, back hygiene counselling and postural education at the workplace.

In chronic situations, the therapeutic approach includes the following measures:

- a) drug treatment for the pain, in acute stages;
- b) rest must be avoided;
- c) re-education, through exertion restriction programmes, postural adaptation techniques and vertebral ergonomics;
- d) use of a lumbar support band during actions of exertion.

Local treatment with posterior interapophyseal injection is of interest for cases of arthritis and facet disease. Patients must also be evaluated and provided guidance for social problems arising from the suspension or reduction of work activities, generally associated to depression.

Emergency surgical treatment is determined by the neurological condition. The following are indications of decompression emergencies:

- a) cauda equina syndrome
- b) paralysing and/or hyperalgetic lumbar sciatica;
- c) progressive neurological deficits;
- d) persistent neuromotor deficit.

Decompression and/or stabilising surgery (intervertebral arthrodesis), excluding the emergency situation, depends on a careful morphological and functional analysis, thereby rigorously establishing the anatomical-clinical and clinical-radiological correlations. The patient's knowledgeable and informed participation in the decision for surgery is important due to inconsistent results in the medium and long term.

In cases of low back pain associated to lumbar spinal stenosis, the treatment is conservative and surgery should be reserved only in cases of serious neurological compromise.

## FOLLOW-UP

Most cases of common low back pain improve:

- a) over 50% of patients improve after 1 week;
- b) over 90% improve after 8 weeks.

Nevertheless, in 7% to 10% of cases, the symptoms remain for over 6 months. These are the main cases responsible for the high social and economic cost of low back pain. There is recurrence in 50% of cases in the first year after the initial crisis. Each new episode increases the probability of a recurrence and of progression to a chronic condition.

Radicular pain may complicate the course and alter the prognosis of lower back pain.

Non-common low back pain has variable courses and prognoses, depending on its etiology.

Follow-up of patients with low back pain is essentially clinical and has the following goals:

- a) to identify symptoms indicating an uncommon etiology;
- b) to detect whether surgery is necessary due to a deterioration of the initial neurological condition.

The following are signs of uncommon low back pain:

- a) pain begins insidiously and progressively, without a triggering factor, especially if the patient is over age 50 and without a background of low back pain;
- b) atypical topography and characteristics of pain;
- c) increasing pain intensity;
- d) stiffness in the lumbar spine;
- e) alteration of general condition such as asthenia, anorexia, weight loss or fever;
- f) a suspicious history, such as tumours, epidemiology favouring infectious diseases or drug addiction.

The presence of any one of these factors calls for complementary diagnosis exams.

The following are factors entailing a risk of development into a chronic condition:

- a) age;
- b) duration of leave from work;
- c) number of relapses;
- d) need for hospitalisation;
- e) low educational and resource levels;
- f) low self-esteem;
- g) depression and anxiety;
- h) lack of family support and understanding;
- i) poor working conditions;
- j) low job qualifications and work dissatisfaction;
- k) precarious job;
- l) conflict following a work accident;
- m) poor treatment.

#### PATIENT REFERRAL

The initial approach to common low back pain is handled by the primary health providers

Patients are referred to other specialists when a non-common origin is suspected, when it becomes chronic or when there are neurological complaints or suspicion of vertebral instability.

#### PERIARTICULAR RHEUMATIC DISEASES

##### DEFINITION

Periarticular rheumatic diseases (PRD), also called diseases of soft or abarticular tissue, are a broad group of musculoskeletal pain syndromes that

result from pathology of bursae, tendons and their synovial sheaths, entheses, muscles and fasciae.

They are clinically characterised by localised pain in juxta-articular sites, aggravated by certain movements, which distinguishes them from joint disorders. However, occasionally it may be necessary to perform specific manoeuvres in the physical exam, to determine the extra-articular nature of the pain's origin.

PRD are responsible for a large number of consultations both in primary care and hospitals.

PRD may be classified topographically as localised or multiple:

- a) according to the etiology and pathogenesis, such as injuries caused by repetitive movements;
- b) according to a clinical and pathological description, such as tendonitis or bursitis.

## RISK FACTORS

This pathological group is associated with certain occupational, sports, or recreational activities.

Most PRD associated to a work-related activity are included in the occupational musculoskeletal injuries. However, regardless of the circumstances under which PRD emerge, people potentially most affected are those who chronically suffer from low impact and repetitive trauma.

## PREVENTION

Early diagnosis of PRD depends on pain characteristics and on the disability it causes. Therefore, an adequate evaluation of symptoms, of the circumstances in which they appear and of their modulation, is essential and must be complemented with a systematic physical exam. This is relevant because PRD may occur alone or in association with systemic diseases such as rheumatoid arthritis, which hampers the diagnosis and requires specific therapy.

Early diagnosis and adequate therapeutic interventions are factors that influence the passage to chronicity, especially in the workplace, where causal factors persist.

The prevention of PRD depends on the knowledge and correction of the causes. Generally, in situations related with sports and leisure, prevention calls for complying with general measures such as:

- a) selecting an adequate sport;
- b) selecting appropriate equipment;
- c) planning an exercise programme according to the individual's anthropomorphic characteristics.

#### TREATMENT

Most PRD are self-limited, improve within weeks, and only some patients will maintain symptoms for more than 6 months.

Since the clinical entities included under this heading are heterogeneous, it is not possible to recommend a universal treatment for PRD. On the other hand, there is no scientific evidence on what is the most effective approach in most of these situations.

The goals of PRD treatments include:

- a) pain relief;
- b) reduction of disability.

Generally, treatment should include:

- a) correction of the identified causes;
- b) medication use;
- c) early rehabilitation.

Medication include analgesics, NSAIDs that are either systemic or transdermic and/or the use of local corticosteroid injections with or without local anaesthetics.

## FOLLOW-UP

Generally, PRD are not serious but – due to their frequency, reoccurrence and resulting disability – may be a major cause of professional absenteeism and of personal suffering. Much of that suffering also arises from the difficulty in diagnosing the problem and subsequently from the delay or inadequate treatment.

However, an adequate approach, including the correction of all factors involved in the cause and maintenance of PRD leads to a quick recovery without consequences for a cure.

PRD are diseases that are generally self-limited and curable and thus, as a rule, do not require follow-up.

## PATIENT REFERRAL

Most PRD must be identified and treated through primary health care, at times through a physiatrist. However, if there is no obvious clinical improvement or when a systemic rheumatic disease is suspected, the patient must be referred to rheumatology or to orthopaedics.

## WORK-RELATED MUSCULOSKELETAL INJURIES

### DEFINITION

Work-related musculoskeletal injuries (WRMI) are a broad and diverse group of disorders that in most cases overlap PRD and differ from these by including some situations of osteo-articular injuries and of synovium sacs and their common association to occupational risks. The symptoms of WRMI are generally similar to those of PRD.

Since the consequences of WRMI are disability, loss of productivity and personal suffering, we may consider them as one of the main problems of occupational medicine.

WRMI are grouped into three categories:

- a) injuries of tendons and their sheaths;
- b) nerve injuries;
- c) neuro-vascular injuries.

### RISK FACTORS

Risk factors related with WRMI may be of an ergonomic, organisational or individual cause.

The following risk factors are of an ergonomic origin:

- a) repetitive movements requiring strain;
- b) mechanical shock;
- c) grasping force and palm load;
- d) static and muscular load;
- e) mechanical stress;
- f) vibration and extreme temperature;
- g) unadequate positions that may be caused by poorly designed equipment, by tools or the workstation.

The following risk factors are of an organisational origin:

- a) excessive work hours and pace;
- b) work with an imposed work pace (assembly lines);
- c) insufficient pauses or rest from work;
- d) lack of work safety or dissatisfied workers;
- e) excessive supervision, such as video monitoring.

The following risk factors are of an individual origin:

- a) smoking;
- b) excessive intake of alcoholic drinks;
- c) obesity.

A higher number of risk factors increase the risk of developing WRMI. One of the main risk factors for the development of tendonitis and other WRMI is repetitive work, since the risk is closely related with the repetition frequency, either in itself or in association with other risk factors.

### PREVENTION

Preventative measures for WRMI must focus not merely on the individual but also on the workplace. The workplace and tools must be adapted and mechanisms must be implemented to compensate for repetitive movements, vibration and inappropriate posture.

Due to the high rate of recurrence, in some cases consideration must be given to assigning other tasks to the worker.

### TREATMENT

The treatment of WRMI coincides with the treatment of PRD, although it must be complimented by intervention at the workplace.

### FOLLOW-UP

Since there is no uniform rule about the intervals and type of follow-up, it must be adapted to each case, depending on the severity of the situation.

### PATIENT REFERRAL

WRMI requires that patients be referred to occupational medicine. Referring patients to rheumatology is mandatory in prolonged cases or in cases that become rapidly disabling. Patient referral to other specialties must be determined case by case.

## OSTEOPOROSIS

### DEFINITION

Osteoporosis (OP) is a systemic skeletal disease characterized by reduction in bone mass and of the quality of the bone's micro-structural quality, that lead

to a decreased bone resistance and a consequent increase of the risk of fractures, which occur more commonly in dorsal and lumbar vertebrae in the distal radius and in the proximal femur.

### RISK FACTORS

Primary OP affects essentially:

- a) post-menopausal women;
- b) the elderly, both men and women.

One in three women and one in eight men over 50 are affected by OP.

The following are regarded as non-modifiable risk factors of OP:

- a) Female gender;
- b) Age over 65;
- c) Caucasian and Asian race;
- d) Positive family history of fracture.

The following are potentially modifiable risks of OP:

- a) early menopause;
- b) hypogonadism;
- c) periods of prolonged amenorrhea;
- d) low body mass index ( $< 19 \text{ kg/m}^2$ );
- e) prolonged immobilisation;
- f) diseases that alter bone metabolism, such as endocrine disorders, chronic rheumatic diseases, renal failure or anorexia nervosa;
- g) intake of drugs that decrease bone mass, such as corticosteroids, anticonvulsive agents and anticoagulants;
- h) lifestyle, such as diets low in calcium, sedentary habits, smoking, alcoholism and excessive consumption of caffeine.

In addition to the factors that reduce bone mass, there are independent factors that facilitate the occurrence of fractures:

- a) a previous fracture;
- b) maternal history of osteoporotic fracture;
- c) length of the femoral axis;
- d) slender build;
- e) cognitive, visual and hearing impairment.

Since fractures caused by fragility normally result from falls, it is essential to know the underlying risk factors:

- a) prior history of falls;
- b) poor physical shape, with alterations in walking and reduced quadriceps strength;
- c) excessive alcohol consumption;
- d) use of drugs such as antidepressants, sedatives and/or antihypertensive agents.

#### PREVENTION

Early diagnosis of OP is determined by measuring the bone density through dual-energy X-ray absorptiometry – DEXA – which may be used to identify three diagnostic categories:

- a) normal (T score greater than -1);
- b) osteopenia (T score between -1 and -2.5);
- c) osteoporosis (T score less than -2.5).

This method is also of prognostic value, to determine the risk of fracture, where the risk is doubled for each decrease in one standard deviation. Although the DEXA has a high rate of specificity and sensitivity, a strategy of universal screening does not have a feasible cost-effective rate for its applicability to the general population. It is necessary to carefully identify individuals under risk, a task to be performed by primary health care services through the clinical history and by applying risk rates and/or standards to

evaluate the need to perform a DEXA. As such, it will be possible to increase the technique's productivity, to sensibly use available resources and to identify individuals who will benefit the most from therapeutic intervention.

The guidelines defined by the WHO for performing DEXA are:

- a) hypogonadism (early menopause, prolonged secondary amenorrhea, primary or secondary hypogonadism in both sexes);
- b) chronic diseases associated to osteoporosis;
- c) presence of risk factors (maternal history of hip fracture, low body mass index);
- d) prior fracture caused by fragility;
- e) prolonged treatment with corticosteroids;
- f) x-rays showing deformity and/or vertebral osteopenia;
- g) loss of height and/or dorsal cyphosis. (after x-ray confirmation of a vertebral deformity).

Perimenopause or menopause, in themselves, are not indications for measuring bone mass. The measurement must be requested when there is also a history of hip fracture and a low body mass index.

If there is OP, the following must be included in the primary health care approach:

- a) minimal laboratory evaluation, only to identify the most common secondary causes of OP or other causes of fracture;
- b) lateral radiograph of the dorsal and lumbar spine to detect the presence of a vertebral deformity.

There is still insufficient scientific evidence to justify using bone metabolism biomarkers in the OP diagnostic approach.

Primary prevention of OP aims to obtain a good peak bone mass and is based on early detection and correction of the modifiable risk factors, especially factors related with lifestyle (e.g. diet, physical activity).

Secondary prevention must overcome bone loss, particularly in women after menopause and already subject to osteopenia. Besides correcting any risk factors that are still modifiable, it may be necessary to use pharmacological measures (e.g. hormonal replacement therapy, bisphosphonates – alendronate and risedronate – raloxifen, calcium supplements and vitamin D). Pharmacological measures should be considered when the T score is below -2.0, associated to major risk factors for osteoporotic fractures.

Tertiary prevention, when OP already exists, is meant to prevent fragility-induced fractures. In that situation, in addition to pharmacological and non-pharmacological measures, it is essential to evaluate and correct risk factors for falls and other trauma, even when they are minimal.

## TREATMENT

Persons with a history of fragility fractures, even if the DEXA reveals only osteopenia, or persons for which a justified DEXA revealed a T-score of less than -2.5 must be given OP treatment. The therapeutic approach must include non-pharmacological and pharmacological measures.

In very old persons who are institutionalised or with limited mobility and at risk for falls the following measures should be considered:

- a) intake of calcium and vitamin D supplements;
- b) use of hip protectors;
- c) fall prevention.

## FOLLOW-UP

In most cases, OP evolves asymptotically, and has good prognosis if correct therapeutic intervention is applied. On the contrary, serious OP (causing fractures) has greater morbidity and mortality.

Patients with vertebral fractures, even when subject to therapy, have various medical complications, lower quality of life, psychopathological alterations and lower life expectancy.

Patients with hip fractures have a mortality rate of 20% to 30% within one year of the fracture. Only 15% of patients with a hip fracture recover their prior functional capacity and 40% are left with a serious disability.

Periodic follow-up of osteoporotic patients is, essentially, clinical to determine whether new fractures have occurred or, in the case of prior vertebral fractures, to detect complications resulting from the fracture such as back pain, respiratory failure or gastrointestinal problems.

Due to the fact that about 2/3 of vertebral fractures are asymptomatic, it is essential that the physical exam include a measure of the patient's height and the evaluation of dorsal kyphosis.

There are no specified criteria for periodic x-rays to the dorso-lumbar spine. These exams should only be performed if a vertebral fracture is suspected.

There is no need for regular laboratorial evaluation of osteoporotic patients.

Women on hormonal replacement therapy must be subject to a gynaecological and breast examination at least once a year.

The interval between a new bone mass measurement depends on the initial reading and on the patient's age. Persons over age 65 and with a first DEXA do not need to repeat the exam.

Post-menopausal women with a normal reading in the first justified DEXA may repeat the exam after the age of 65.

In patients undergoing therapy, DEXA should not be repeated before 18 to 24 months of proper treatment. When a first evaluation reveals osteopenia, the decision to repeat the exam must be individual, depending on the patient's age and on the T-score, but never before 2 years have elapsed.

Just as menopause does not justify measuring bone mass, there is also no justification for a post-menopausal woman to repeat DEXA when the aforementioned criteria are not met.

#### PATIENT REFERRAL

Primary health care is responsible for the prevention and treatment of OP.

Patient referral to rheumatology is justified:

- a) in all cases of OP fractures;
- b) in glucocorticoid-induced OP;
- c) in secondary OP;
- d) in all cases where, despite proper treatment, bone mass continues to decrease.

OP secondary to endocrine causes and male OP must be referred to endocrinology and rheumatology.

## FIBROMYALGIA

### DEFINITION

Fibromyalgia (FM) is a rheumatic disease of unknown cause and of functional nature which causes diffuse pain in the soft tissues, whether they are muscles, ligaments or tendons, but it does not affect joints or bone.

The pain caused by FM is followed by quantitative and qualitative sleep disturbance, fatigue, headache and cognitive alterations such as memory loss and difficulty in concentration, paresthesias / dysesthesia, irritability and, in about 1/3 of cases, depression.

### RISK FACTORS

FM affects about 2% of the adult population. The following are risk factors:

- a) gender (women are 5 to 9 times more affected than men);
- b) age (starts between 20 and 50 years of age)

Children and adolescents may also suffer from FM, although the rate is the same in boys and girls of school age.

### PREVENTION

The diagnosis of FM is essentially clinical, and complimentary tests are used to exclude other diseases with similar symptoms.

The diagnosis of FM is based on the presence of:

- a) general musculoskeletal pain, that is, below and above the waist and in the left and right halves of the body;
- b) pain lasting more than three months;
- c) the presence of painful points on applied pressure in symmetric areas of the body and well established locations.

Although it is necessary to have at least 11 tender points, out of 18 potential ones, to classify this syndrome, that number may not be necessary to establish the diagnosis.

The diagnosis must distinguish cases of inflammatory rheumatic disease, thyroid dysfunction and muscular pathology.

There are no primary prevention standards for FM. However, there is knowledge of the risk factors associated to states of generalized chronic pain:

- a) female gender;
- b) age between 40 and 60;
- c) low income level;
- d) low educational level;
- e) divorced/separated.

Characteristics of personalities prone to pain are also known:

- a) dedicated workers;
- b) excessively active persons;
- c) compulsive perfectionists;
- d) incapacity to relax and enjoy life;
- e) negation of emotional and interpersonal conflicts;
- f) incapacity to deal with hostile situations;
- g) need for affection;
- h) child-like dependence.

Warning signs for the development of FM have also been determined:

- a) family history of the disease;
- b) prior painful syndrome;
- c) concern with the prognosis of other coexisting diseases;
- d) vertebral trauma, especially of the cervical spine;
- e) incapacity to deal with adversities;
- f) history of depression/anxiety;
- g) persistent symptoms of "virosis;"
- h) sleep disturbance;
- i) substantial emotional dysfunction;
- j) work-related pain.

Knowledge of these warning signs allows for early intervention and secondary and tertiary prevention, thereby preventing FM from deteriorating and complications from developing. The success of these actions depends, however, on whether primary health care providers know and give importance to the factors associated to, preceding and accompanying FM, as well as those that deteriorate its prognosis.

## TREATMENT

FM must be treated in primary health care.

After the diagnosis, an explanation of the nature of the disease is crucial for its proper treatment. The patient must also be informed on factors improving and deteriorating the condition and on the normally good prognosis of the disease. The patient must also be educated on lifestyle, the practice and type of exercise and relaxation techniques.

Drugs prescribed with the greatest efficacy are analgesics, tricyclic antidepressants and selective serotonin reuptake inhibitors, muscular relaxants and sleeping pills.

Regular exercise is recommended.

Other therapeutic means, as well as intervention by rheumatology, psychiatry and other medical specialties or of other non-medical health care providers are frequently necessary.

#### FOLLOW-UP

FM does not cause deform or permanent physical disability, but many patients do not tolerate medication and, among those who tolerate it, less than 50% show significant improvement.

In specialised centres, the treatment's success rate may reach 60%.

Professions requiring workers to maintain the same posture for long periods, repetitive movements and frequent and/or maintained elevation of the upper limbs are the ones most difficult to tolerate.

Experience shows that, generally, patients who retired early due to FM get worse because of the following factors:

- a) less physical activity;
- b) less time spent in distraction environments;
- c) greater depression;
- d) feeling useless;
- e) more time to think about the disease.

Patients suffering from FM must be evaluated periodically for their complaints and for any adverse effects related to therapy.

There are no remission criteria for the disease but, in view of the treatment objectives, the pain must be evaluated with validated instruments.

It is essential to systematically compare the following parameters in comparison with prior evaluations:

- a) level of activity;
- b) perfectionism;
- c) assertiveness;

- d) capacity to deal with obstacles and stress;
- e) sensitivity to pain;
- f) depression;
- g) sleep disturbance;
- h) anxiety/anguish.

It may be advantageous to obtain some of this information from people who interact directly with the patient.

Follow-up intervals will depend on the severity of FM and on the characteristics of associated conditions.

#### PATIENT REFERRAL

About 60% to 70% of patients with FM do not improve with therapy or did not tolerate medication, at times to a great extent. In these cases, the patient should be referred to rheumatology for a re-evaluation and possible therapy guidance.

## MICROCRYSTAL-RELATED ARTHROPATHIES

### DEFINITION

Crystal-Related arthropathies are a group of diseases whose clinical manifestations and pathological alterations are caused by the deposition of mineral crystals in musculoskeletal tissue. Gout is a disease resulting from the deposition of monosodium urate crystals in the tissue due to the supersaturation of extracellular fluids in uric acid. The situation may be asymptomatic or cause acute or chronic arthritis, gouty tophi, nephropathy with renal failure and nephrolithiasis.

The disease is caused by the deposition of calcium pyrophosphate dihydrate crystals and it is characterised by the deposition of those crystals in the joint cartilage and joint fibrocartilage (radiographic chondrocalcinosis). Most commonly asymptomatic, consisting of a radiographic finding, it may cause

acute arthritis (pseudogout) more frequently in the knee. In rare cases, it may cause serious forms, hemarthrosic, in particular affecting the knee and the shoulder, and rapidly destructive arthropathy. In rare cases, patients develop a chronic symmetric polyarthritis that course with inflammatory bouts of moderate intensity that may simulate rheumatoid arthritis. Its association with osteoarthritis is not clear, but it should be suspected when radiographic signs of serious osteoarthritis are found and/or when found in unusual sites.

Hydroxyapatite crystals are, in most cases, associated to tendon and/or bursae calcifications. In most cases, they are asymptomatic, but there may be a partial resolution of the deposit, resulting in acute periarticular inflammation. In rare cases, these calcifications involve multiple tendons and/or bursae, revealing a condition of multiple tendon calcifications. In rare cases, the intra-articular deposit of hydroxyapatite crystals may be the cause of rapidly destructive arthropathy, with bloody synovial fluid effusion.

#### RISK FACTORS

Gout results from hyperuricemia, and the latter may be caused by an overproduction of uric acid or its underexcretion.

The overproduction of uric acid may result from a new increase in purine synthesis, from excess dietary purine consumption, or from a variety of diseases in which there is an increase in the cellular nucleotide turnover, with the consequent release of metabolised nucleic acids into urate such as seen in myeloproliferative and lymphoproliferative disorders. Even rarer, are hereditary abnormalities of purine metabolism associated with early and serious forms of gout.

Excessive alcohol consumption may cause overproduction of urate, but also decreases the renal excretion of uric acid, since alcohol increases the production of lactic acid. Most patients with hyperuricemia are underexcretors of uric acid, more commonly idiopathic, caused by deficient secretion or increased tubular resorption. However, various other causes have been identified, such as:

- a) renal failure;
- b) dehydration;
- c) acidotic states;

- d) high blood pressure;
- e) hyperparathyroidism;
- f) use of drugs, such as diuretics, cyclosporine A, pyrazinamide, ethambutol and low-dose aspirin.

Chronic lead poisoning causes a renal tubular disorder associated to the so-called saturnine gout;

Therefore, persons at risk to develop gout are those:

- a) whose diet includes large amounts of purines associated to animal protein consumption;
- b) who drink alcohol in excess;
- c) take drugs such as thiazides and some tuberculostatic agents;
- d) have impaired renal function;
- e) are subject to dehydration or acidosis;
- f) are at risk for exposure to nephrotoxic substances such as lead;
- g) suffer from haematological diseases that cause excessive cellular production.

Primary gout is much more common in men. In women it almost only appears after menopause.

Calcium pyrophosphate dehydrate deposition disease is mostly idiopathic, more common in women and its incidence increases with age.

There are various hereditary forms of the disease, generally with a dominant autosomic transmission, appearing earlier and with a less common pattern of joint involvement, as well as forms associated with metabolic diseases.

Joint surgery and trauma may trigger acute attacks, similar to what happens in gout .

Forms of arthropathy and periarthropathy associated to the deposition of hydroxyapatite crystals appear more frequently in persons after the age of

fifty and in structures suffering from some degree of injury, often caused by chronic or repetitive trauma.

### PREVENTION

The definitive diagnosis of a crystal-related arthropathy depends on the identification of the typical crystals. These crystals may be identified in the synovial fluid through optical microscopy with ordinary light, which identifies the type of crystal by the shape, or under the effect of polarised light by which its birefringence is observed. The hydroxyapatite crystals are identified by colouring the synovial fluid with alizarin-S red or by electronic microscopy.

Early diagnosis is important in gout because adequate treatment, including the control of hyperuricemia is effective in preventing the complications of disease.

In diseases caused by other crystals, typical calcifications are more often found in asymptomatic individuals and it is not possible to identify or to forecast which one will show clinical manifestations.

Prevention of gout is based on identifying hyperuricemia and its treatment. There is consensus that therapeutic intervention is necessary when a reading of over 11 mg/dl of uricemia is found.

Patients must avoid excessive purine-rich foods and alcohol and also avoid other modifiable risk factors such as some drugs, in particular diuretics, nephrotoxic agents and diseases with repercussion on kidneys.

Patients with myeloproliferative and lymphoproliferative disorders taking cytostatic drugs must be treated preventatively with allopurinol, and dehydration and acidotic states must be avoided.

Secondary prevention in gout calls for controlling uricemia in patients that have already had at least one acute attack of arthritis and/or nephrolithiasis due to urates, and for reinforcing hypouricemic treatment prior to surgery, in serious disease or trauma. Intercritical treatment with low doses of colchicine prevents acute arthritis attacks.

In tertiary prevention, it is important to avoid obstructive uropathy and uric nephropathy by maintaining a high diuresis through abundant water intake and urine alkalization to prevent the formation of calculi. Also included is surgery for removing gouty tophi and the treatment of joint complications.

In calcium pyrophosphate dehydrate deposition disease, prevention is based on diagnosis and treatment of the metabolic conditions that may cause the disease.

## TREATMENT

In gout, a distinction must be made between treatment and prevention of acute attacks of arthritis, by the application of colchicine or NSAIDs, and the underlying hypouricemic treatment.

Treatment of gout is based on general measures and hypouricemic treatment. The former include weight loss in obese patients, treatment of associated metabolic conditions (e.g. diabetes, hyperlipidemia), decreased intake of purine-rich foods and alcohol.

Nephrolithiasis and urate nephropathy are prevented by the intake of alkaline water to ensure an abundant diuresis:

Hypouricemic treatment is necessary when:

- a) hyperuricemia is greater than 11 mg/dl;
- b) there are gouty attacks;
- c) gouty tophi;
- d) nephropathy;
- e) nephrolithiasis.

Hypouricemic treatment is based on allopurinol to attain uricemia below 5 mg/dl. Patients who cannot tolerate allopurinol must be treated with uricosuric drugs.

Treating of calcium pyrophosphate deposition disease is symptomatic.

Periarticular RD, such as tendonitis and bursitis, associated to the deposition of hydroxyapatite crystals, are treated with rest and, possibly, local corticosteroid injections.

#### FOLLOW-UP

Initial attacks of gout generally improve spontaneously in less than one week. However, if it is not properly treated, episodes become more frequent, affect more joints and progress to a chronic polyarticular disease, with a permanent state of pain and substantial disability. Gouty tophi and/or complications of the urinary system may also occur.

As for calcium pyrophosphate dehydrate deposition and hydroxyapatite crystal deposition diseases, the hereditary forms and those associated with metabolic diseases progress if the cause is not treated or treatable. The primary forms have a fluctuating course, with intervals of variable duration between attacks, but are often uniform for each patient, and incapacitating forms are rare. A joint diagnosis of rapidly destructive forms is very poor, and it is almost always necessary for the affected joints to be replaced by a prostheses.

Monitoring of patients with gout is, particularly, essential until it is ensured that the joint symptoms, either acute or chronic, have eased and the uricemia has been stabilised below 5 mg/dl. This stability is important since the attacks are frequently provoked by fluctuations of uricemia, even uricemia does not reach very high values. Once this stage has been overcome, monitoring the patient every 6 months may be sufficient.

The other crystal-related arthropathies do not need specific monitoring.

#### PATIENT REFERRAL

Cases of gout must be referred to rheumatology when there are doubts in the diagnosis, either of the acute or chronic forms, at times only overcome by arthrocentesis to identify crystals in the synovial fluid. Another reason for patient referral is the difficulty in treating, either due to ineffectiveness or intolerance.

The other crystal-related arthropathies must also be referred when there are doubts about the diagnosis that may require identifying the crystals or the need for an injection, arthrocentesis or flushing of the joint.

Treatment of periarticular RD implying local injections requires referral to rheumatology or to orthopaedics.

## RHEUMATOID ARTHRITIS

### DEFINITION

Rheumatoid arthritis (RA) is a chronic inflammatory disease of unknown origin, that may occur at all ages. Its predominant presentation is repeated and usually there is chronic involvement of the joint and periarticular structures and it may, however, affect the connective tissue in any part of the body and give rise to a wide variety of systemic manifestations.

Due to its prevalence and the serious personal, economic and social problems it causes RA is, indisputably, the main systemic rheumatic disease.

When RA is not treated early and correctly, as a rule it brings serious consequences to patients, implying functional and work disability, high co-morbidity and increased mortality compared with the general population.

### RISK FACTORS

Females are affected more frequently by RA, in a proportion that, in Portugal, is of 4:1.

Although RA may emerge at all ages, even in children and the elderly, it is mainly a disease of young adults and of pre-menopausal women.

Excluding age and gender, the other risk factors, such as blood transfusions prior to the start of RA, obesity and smoking are controversial and seem to be more related with a worse progression and prognosis of the disease than in the risk of contracting it. Some cases of RA appear sporadically after infections by parvovirus and by rubella, tetanus, hepatitis B and the flu.

Among possible protective factors, emphasis goes to being male, pregnancy, oral contraceptives and the moderate intake of alcohol.

### PREVENTION

Early diagnosis of RA is essential. When this disease is diagnosed in the first three to six months of its clinical development and is correctly treated, there is a high probability of impeding functional and work disabilities, of decreasing co-morbidity and of maintaining average life expectancy.

Diagnosing RA is a medical urgency, since 90% of patients have bone erosions at the end of 2 years if they are not diagnosed and treated early and correctly.

Clinically, RA may begin as an acute polyarthritis, at times with fever, or by a monoarthritis, by a tenosynovitis and, most of the times, insidiously by small joints of the hands and feet. Exceptionally, the presenting symptom may be a systemic manifestation.

The joints most often and earlier affected by RA are the 2<sup>nd</sup> and 3<sup>rd</sup> metacarpophalangeal joints, the proximal interphalangeal joints of the hand and the metatarsophalangeal joints.

Patients with polyarthritis or suspicion of having RA, must ideally be observed by a rheumatologist in the first ten weeks of symptom appearance, regardless of having criteria for the diagnosis of the disease.

The early diagnosis of RA is based on the presence of:

- a) swelling of three or more joints;
- b) involvement of the metacarpophalangeal and/or metatarsophalangeal joints;
- c) morning stiffness of more than thirty minutes;
- d) symmetry of joint involvement.

The early diagnosis of RA must not, however, be established without 12 weeks having elapsed after the start of symptoms.

Laboratory tests and conventional radiography are not pertinent for early RA diagnosis. However, early diagnosis may be made through bone scanning, ultrasound and magnetic resonance imaging that reveal synovitis after only a few weeks.

The prevention of RA may be secondary or tertiary.

Secondary prevention, which aims to decrease disease severity, implies early diagnosis and early and correct treatment.

Tertiary prevention, which aims to reduce functional disability and improve quality of life, may be performed by patient education, weight control and physical activity, surgery and rehabilitation.

For patients who are serious motor-impaired, the following is important:

- a) environmental improvement;
- b) professional training, orientation and rehabilitation;
- c) protected employment;
- d) home assistance.

## TREATMENT

In the past 15 years, the treatment of RA has undergone profound changes due to the evaluation of inflammatory activity and the enhanced knowledge of factors underlying worse diagnoses, the early use of slow-acting antirheumatic drugs, the emergence of combined therapy and, in the previous years, the application of biologic therapy.

The purpose of RA treatment is the remission of diseases that, when not obtained, is considered a therapeutic failure. The main therapeutic strategies are:

- a) patient education;
- b) general measures, including rest;
- c) exercise;
- d) psychological support;

- e) drug therapy that, in addition to fighting pain and inflammation, must prevent and treat associated diseases;
- f) surgery.

#### FOLLOW-UP

The prognosis of RA is restricted by various factors, and it is worse when there are:

- a) a large number of swollen joints;
- b) rapidly progressive disease;
- c) systemic injuries;
- d) low education;
- e) persistently elevated acute phase proteins;
- f) presence of rheumatoid factor in the serum;
- g) presence of histocompatibility DR4, DRB1 antigens and of the epitope;
- h) early erosions;
- i) high rate of functional disability (Health Assessment Questionnaire);
- j) early joint replacement surgery.

RA evaluation is very important, since its treatment and its follow-up are limited by the disease's inflammatory activity, by measuring functional capacity and factors associated to worse prognosis.

The inflammatory action is measured using indexes obtained through the number of swollen joints, the number of painful joints, erythrocyte sedimentation rate, physician and patient global assessment of the disease, pain and functional capacity severity.

After the initial diagnosis, patients must be monitored on a monthly basis until establishing a therapy that controls the disease's inflammatory activity and, then, every six weeks and every three months.

To monitor these patients, it is necessary to determine, at least, in each consultation, the inflammatory activity of RA, to evaluate it in the laboratory and to determine whether there are side effects from the various prescribed drugs.

#### PATIENT REFERRAL

All patients with RA must be referred to rheumatology.

### SERONEGATIVE SPONDYLOARTHROPATHIES

#### DEFINITION

Seronegative spondyloarthropathies (SNSA) are a group of chronic rheumatic diseases that share some common clinical manifestations such as the involvement of the axial skeleton, asymmetric peripheral arthritis, enthesopathy and mucocutaneous manifestations and some typical laboratorial findings such as high prevalence of the histocompatibility antigen HLA-B27 and the lack of rheumatoid factors.

This is a group of multisystemic diseases characterised by inflammation of the spine, peripheral joints and periarticular structures.

The designation includes a group of diseases:

- a) ankylosing spondylitis (AS);
- b) Reiter's syndrome (RS);
- c) reactive arthritis (ReA);
- d) psoriatic arthritis (PA);
- e) Spondylitis associated with inflammatory bowel disease (SAIBD).

There are also syndromes that share various characteristics of SNSA but do not comply with criteria to be classified as one of the types defined above, and are designated as undifferentiated spondyloarthropathy (uSpA).

SNSA consist of a group of diseases that share, as their most characteristic element, the inflammatory involvement of the sacroiliac joints and of the lumbar spine, causing loss of mobility and joint stiffness.

### RISK FACTORS

Typically, SNSA appears around the age of 20 for AS, but it is not possible to determine stages of greater risk for all the other SNSA subtypes.

SNSA are described, in particular, as diseases affecting males. Nevertheless, today it is becoming accepted that there may not be a difference between genders.

Despite this controversy, it is still accepted that males are more likely to contract some clinical forms such as AS and juvenile AS, whereas some subtypes equally affect both genders such as ReA, SAIBD and uSpA, or are more common in females, such as PA. One specific case is juvenile AS that, by definition, appears in individuals under 16 years of age and is more common in males.

As a consequence of the importance of genetic susceptibility in the development of these rheumatic diseases, in which there is a prevalence of the HLA-B27 allele in the population of affected individuals that varies from 50% to 95%, there is a tendency for SNSA to affect family clusters, as recognised in various epidemiological contexts.

### PREVENTION

The diagnosis of SNSA is essentially clinical, but its confirmation frequently requires complementary means of diagnosis to which access is relatively easy such as conventional radiography and class I HLA typing. Therefore, in most cases, the diagnosis is evoked only after the appearance of complaints suggesting a diagnosis.

One of the critical problems of early diagnosis of SNSA arises from the fact that there are reservations in making the diagnosis without radiological evidence of sacroiliitis. This reservation is maintained in relation to families of patients with recognised SNSA and when there are clinical characteristics of inflammatory compromise of the spine.

This deficient ascertainment of the disease's presence, even when sufficient clinical elements to establish the diagnosis exist, leads to a situation in which many patients are not diagnosed for several years and, therefore hampering early diagnosis.

In fact, it is acknowledged that a 5-year gap may exist between the first signs and symptoms of the disease and typical radiological features are recognized.

Moreover, some patients may only have thoracic pain and stiffness without radiographic evidence of sacroiliitis or even without inflammatory low back pain, thus causing a further obstacle to recognising the diagnosis.

Early recognition of this group of diseases, or at least, early enough, is an issue to be dealt with by primary care physicians and thereby justifies the assumption that all patients with back pain may have SNSA so that referral can be made in useful time.

Primary prevention of SNSA is still not regarded as possible due to the role played by factors such as genetics, not yet placed within the framework of the disease's etiopathogenesis. Moreover, environmental factors, which are considered equally important, are not subject to specific measures to prevent SNSA since they are not clearly individualised. There are no means of establishing individual or population-based strategies to prevent the disease, nor even among populations selected for their greater probability of contracting the disease, such as relatives of persons with SNSA.

As for secondary prevention, resulting from early or at least timely recognition of the disease, there is an extensive field in which it is possible and desirable to improve diagnostic sensibility, so that individuals, up to now not identified as such, may be recognised as carriers of SNSA. The adoption of proper treatment strategies may, in early stages, determine large differences in the functional prognosis in the medium term.

Tertiary prevention, guided towards correcting established sequelae, even when very specialised, has the least cost-effective results. Moreover, even when only the patient's quality of life is considered, the available invasive and non-invasive techniques are not very effective in inducing significant differences. The exception to this analysis may be joint replacement surgery such as hip replacement. Nonetheless, these are expensive techniques that resolve a localised limitation in diseases in which the compromise is generally multi-focal.

## TREATMENT

Treatment of most patients with SNSA aims to relieve pain, stiffness and fatigue, to maintain adequate posture and good physical and psychological states. Thus, in addition to adequate patient information and education, the therapeutic programme includes physical exercises such as swimming and stretching or regular sports activity.

Early institution of appropriate physiotherapy programmes is also a determinant for the prognosis, with proven efficacy in terms of mobility of the dorsal and lumbar spine and of the general well-being of patients. Besides kinesiotherapy, physical agents and the teaching of postural correction practices may also play an important role.

The prescription of medicines completes the therapeutic arsenal.

Surgery is especially important since it is capable of replacing some major joints that are frequently affected or of correcting fixed spine deformities by performing vertebral osteotomy in serious cases of non-functional positions of ankylosis.

## FOLLOW-UP

Most patients with SNSA:

- a) have a clinical form of the disease that is mild and has good functional prognosis;
- b) never show extraskeletal manifestations, or else those manifestations are limited to anterior uveitis that emerges in about 30% of cases;
- c) have a clinical development characterised by spontaneous exacerbations and remissions;
- d) maintain good functional capacity and are able to continue working even with very advanced forms.

There is no evidence that life expectancy is affected.

The functional prognosis was recently improved by the possibility of performing hip Arthroplasty which resolves one of the main limitations affecting patients with SNSA.

The follow-up of patients with SNSA has not been established, and there is no consensus on the most relevant measures.

#### PATIENT REFERRAL

All patients diagnosed with SNSA must be referred to rheumatology.

## SYSTEMIC RHEUMATIC DISEASES

#### DEFINITION

This designation includes a variety of diseases and syndromes that share identical pathogenic features and clinical findings of diffuse inflammation.

The designation of conjunctive or connective diseases, or of connective tissue diseases, as they are also commonly known, is not etiopathologically appropriate. Using the term autoimmune diseases restricted merely to this nosological group is vague and reductive, since there are autoimmune diseases that affect all organs and systems.

The almost systematic involvement of the musculoskeletal system, such as joints, muscles and bones, associated to frequent involvement of other organs and systems justify the use of the term systemic rheumatic diseases (SRD).

The intimate cause of these clinical conditions is not known, but it is presumed that it results from an interaction between genetic factors and unidentified environmental agents.

In SRD there is a defect in the immune system that is associated to the presence of antinuclear antibodies and other autoantibodies in serum and of inflammatory lesions in various organs and systems that vary according to the disease.

### RISK FACTORS

SRD are of low prevalence, but they are important because of their pathophysiological complexity, their clinical multidisciplinary, the diagnostic difficulty, the diversity of treatments and the prognostic variability.

All SRD are more frequent in women. Some vasculitis are, however, more frequent in men, such as Behçet's syndrome, polyarteritis nodosa and Wegener's granulomatosis.

SRD are ailments that usually begin in young adults, between age 30 and 40, although some vasculitis, such as Kawasaki disease and Schonlein-Henoch purpura and some of the inflammatory myopathies, occur in children and adolescents. Giant cell arteritis begins in individuals over 65 years of age.

### PREVENTION

SRD form a very heterogeneous group, with few common characteristics in terms of their presentation, clinical course and treatment.

It is not difficult to make a specific diagnosis of these diseases when they have various typical signs and symptoms but may be very problematic when they present as a mono or oligosymptomatic form or when complaints appear separated by longer intervals.

Diagnosis, whether early or late, is based on clinical findings, although laboratory or other complementary diagnostic exams have relative importance.

The initial symptoms of these ailments are generally vague and only rarely immediately suggest the diagnosis.

In most patients, SRD present as:

- a) discrete musculoskeletal complaints such as polyarthralgia, pain and/or muscular weakness;
- b) systemic symptoms such as fever, anorexia, malaise, fatigue and adynamia.

Under this atypical presentation, it is difficult, if not impossible, to determine if we are dealing with a self-limited process, a SRD or any other disease.

In rare occasions, SRD are very serious at onset, requiring fast patient referral for treatment. In these cases, the diagnosis is more easily recognised because of its systemic signs and symptoms with high fever, at times with spikes, rapid weight loss and manifestations caused by compromise of an organ or system.

DRS must be regarded as a diagnostic possibility when a young woman has:

- a) compromise of general state;
- b) fever;
- c) polyarthralgia / polyarthritis;
- d) organ involvement, with pericardial and/or pleural effusion;
- e) cutaneous lesion, particularly in the face;
- f) Raynaud's syndrome.

If this clinical evidence is supported by analytical changes, such as high erythrocyte sedimentation rate, proteinuria, polyclonal gammopathy and haematological changes such as leukopenia, neutropenia and lymphocytosis, thrombocytopenia and haemolytic anaemia, then the diagnosis is more probable.

Since the origin of SRD is unknown, primary prevention of these diseases is not possible. Secondary prevention and tertiary prevention are based on early diagnosis and timely and correct therapy, the only way to avoid or minimise the various complications arising from SRD.

## TREATMENT

Treating a group of ailments as polymorphic as SRD will be obviously different according to the condition.

SRD are treated with medicines according to the disease's activity level, severity and clinical manifestations.

## FOLLOW-UP

SRD are potentially serious, with mortality rates that vary, according to:

- a) the level and duration of their activity;

- b) the presence and severity of the lesion of the organ or system or, even, of a multi-organ ailment;
- c) the occurrence of complications such as infection, cardiovascular disease, neoplasia;
- d) the frequent and not rarely serious side effects of aggressive therapy, which is often necessary.

In all SRD, the prognosis is more favourable if diagnosis is made early and therapy is appropriate, greatly assisted by good collaboration between rheumatology and primary care.

Monitoring of SRD must include the following:

- a) analysing the disease's activity;
- b) determining the level of lesion of the target organ/organs;
- c) evaluating tolerance to treatment, particularly side effects of the respective drugs.

For each SRD and category of drugs, the symptoms and clinical signs, as well as the complementary diagnostic exams to be used in follow-up, will be different.

The activity of the diseases, in view of their inflammatory nature, may in most cases be evaluated through the acute phase reactants and of the haemoglobin levels.

#### PATIENT REFERRAL

Taking into account the quantity and heterogeneity of SRD, patient referral criteria cannot be standardised.

However, the following criteria may be individualised for some SRD:

- a) SLE: Patients with mild and stable disease not affecting a target organ, such as the kidney, central nervous system, cardiovascular system and blood, may be monitored, after a confirmed diagnosis by rheumatology, in primary health care at one-month or two-month intervals. The patient must, however, be monitored every half year or every year by

rheumatology. Patients with substantial renal, neuropsychiatric, respiratory and dermatological lupus and with infection, as well as lupus patients who are pregnant must also be monitored by the respective specialists.

- b) SS: Most patients with this syndrome must be monitored in primary health care. The patient must be subject to rheumatology to confirm the diagnosis and when the disease is severe. Ophthalmologic, gynaecological and stomatological care must not be overlooked.
- c) PM/DM: Are very rare and serious conditions that must be monitored by rheumatology and/or dermatology.
- d) SSc: This is an even less common disease than the aforementioned ones. It must be monitored by rheumatology and/or dermatology.
- e) PAN: When this vasculitis is clinically suspected, the patient must be evaluated at least by rheumatology and, if the diagnosis is confirmed, should continue to be monitored there.
- f) WG: Patients with this pathology must be followed by rheumatology and/or pneumology. At times the contribution of nephrology is necessary.
- g) GCA: Patients with GCA must be monitored by rheumatology. It may be necessary to include follow-up by ophthalmology and cardiology.

## JUVENILE CHRONIC ARTHRITIS (JCA)

### DEFINITION

JCA, of an unknown origin consists of a heterogeneous group of diseases initiated before 16 years of age, characterised by the presence of arthritis of one or more joints and lasting for at least 6 weeks.

There is still insufficient knowledge on the pathogenesis of JCA. They are regarded as autoimmune diseases whose etiopathogenesis is multifactorial, involving genetic, immunological and environmental factors.

JCA is classified into different subtypes, according to the clinical and laboratorial characteristics detected in the first 6 months of the disease.

Systemic arthritis – Arthritis with or preceded by daily fever lasting at least two weeks with a daily peak over 39°C, for at least three days, combined with one or more of the following criteria:

- a) brief erythematous evanescent exanthema;
- b) generalised adenomegaly;
- c) hepatomegaly or splenomegaly;
- d) serositis.

Oligoarthritis – Arthritis that affects one to four joints in the disease's first 6 months:

- a) persistent oligoarthritis affects no more than four joints during the disease's whole period;
- b) extended oligoarthritis affects a cumulative total of five or more joints after the disease's first 6 months.

Polyarthritis with negative rheumatoid factors – Arthritis that affects five or more joints during the disease's first 6 months, with persistently negative IgM rheumatoid factors.

Polyarthritis with positive rheumatoid factors – Arthritis that affects five or more joints during the disease's first 6 months, associated to IgM positive rheumatoid factors.

Psoriatic arthritis – Arthritis associated to psoriasis or arthritis and at least two of the following data:

- a) dactylitis, with swelling of one or more fingers, generally in an asymmetric distribution, that extends beyond the joint margin;
- b) onycholysis or nail pitting;
- c) family history of psoriasis in a first-degree relative.

Arthritis related with enthesitis – Arthritis and enthesitis, tender on palpation of the insertion of tendons, ligaments, joint capsules or fasciae on the bone, or arthritis or enthesitis with at least two of the following findings:

- a) tenderness by palpation of the sacroiliac joints and/or inflammatory spondylalgia;

- b) presence of HLA-B27;
- c) family history of the disease associated to HLA-B27, such as ankylosing spondylarthritis, sacroiliitis associated to inflammatory bowel disease or acute anterior uveitis, in a 1<sup>st</sup> or 2<sup>nd</sup> degree-relative;
- d) acute anterior uveitis;
- e) male;
- f) age over 8 at the start of the arthritis.

Other arthritis – Children with arthritis of an unknown cause, lasting six weeks or more and that:

- a) are not classifiable in any of the previous categories;
- b) are classifiable in more than one of the previous categories.

#### RISK FACTORS

No determinant risk factors are known for the onset of JCA.

JCA consist of a group of rheumatic diseases more frequently diagnosed in children, whereby several of them are predominant in females.

The start of JCA before 6 months of age is rare.

#### PREVENTION

To establish a diagnosis of JCA, the presence of the following is necessary: onset of arthritis before age 16, persistence for at least six weeks, and after other children's diseases are excluded, in particular other juvenile arthritis that may simulate one of the subtypes of JCA.

The diagnosis of JCA is clinical, for which there are no specific laboratory exams, although these exams are important for excluding other pathologies. Radiographs in an early stage reveal only swelling of the soft periarticular tissue and juxtaarticular osteopenia. At times, there is periostitis. Arthrocentesis and a biopsy of the synovial membrane may be necessary in monoarthritis or when joint inflammation is out of phase within a context of oligoarthritis or polyarthritis.

Acute anterior uveitis in juvenile spondyloarthropathy is normally symptomatic and therefore easy for the ophthalmologist to diagnose.

Chronic uveitis, found in the oligoarticular form with ANA in the serum is frequently asymptomatic and its early diagnosis needs periodic ophthalmologic exams. Other extraarticular complications that may occur during disease course and that must be detected early are localised or generalised growth alterations, osteoporosis and secondary amyloidosis.

Since JCA is of an unknown origin, there is no primary prevention.

Secondary prevention is based on early diagnosis and adequate early treatment which, in addition to controlling joint inflammation and pain has the following objectives:

- a) prevent and control joint lesions and periarticular structures thereby maintaining or improving their function;
- b) prevent and treat any extraarticular complications;
- c) promote normal physical and psychic growth and development.

Rehabilitation – including occupational therapy and technical aids, orthopaedic surgery, psychological and social counselling to the child and family, education and vocational guidance – is part of the treatment to help limit disability and to help these children and adolescents to be well integrated educationally, socially and professionally.

#### TREATMENT

For successful treatment of JCA, it is necessary to have a multidisciplinary and coordinated team, and it is essential for the family to collaborate.

JCA treatment includes:

- a) education of the child and family;
- b) a number of general measures;
- c) drug therapy;
- d) physical and rehabilitation medicine;

- e) ophthalmologic and stomatological monitoring;
- f) at times, orthopaedic surgery.

It is important to ensure psychological and social support to the patient and family, support to the child's integration at school and his/her early vocational guidance.

General therapeutic measures include:

- a) measures to overcome pain and morning stiffness;
- b) encouragement of correct joint postures and positions;
- c) use of splints to prevent deformities;
- d) daily exercise programme;
- e) balanced diet;
- f) appropriate physical activities;
- g) sports activities;
- h) mouth and dental care.

It is important to prevent and treat any extraarticular complications, in particular uveitis, either acute or chronic, growth alterations and osteoporosis and secondary amyloidosis.

#### FOLLOW-UP

The development of JCA and any side effects of drug therapy must be periodically monitored. Besides monitoring by rheumatology and paediatrics, the patient must be monitored by ophthalmology and stomatology. Periodic laboratory exams are also necessary and, whenever appropriate, radiography of the affected joints.

AJC follow-up must be complemented with a periodic evaluation of the patient's functional capacity and quality of life related with health through the Childhood Health Assessment Questionnaire.

### PATIENT REFERRAL

Children with undetermined arthritis and/or uveitis must be referred to rheumatology.

The diagnosis and follow-up of children with arthritis, particularly JCA, must be performed by a rheumatologist or paediatrician trained and experienced in childhood and juvenile rheumatism.

### EVOLUTION AND PROGNOSIS

The former belief that JCA is generally a benign disease and that most children affected by it reach adulthood without arthritis and without any limitation or deformity has been dispelled in the previous years.

JCA has a very variable and unpredictable prognosis, especially in the disease's initial stages. The prognosis thus depends on the disease's onset and course.

## CLASSIFICATION OF RHEUMATIC DISEASES

### 1. RHEUMATOID ARTHRITIS

### 2. OTHER SYSTEMIC RHEUMATIC DISEASES

2.1. Systemic lupus erythematosus.

2.2. Systemic sclerosis.

2.3. Eosinophilic fasciitis and other variants of systemic sclerosis.

2.4. Polymyositis and dermatomyositis.

2.5. Primary and secondary Sjögren's syndrome.

2.6. Mixed disease of the conjunctive tissue.

- 2.7. Adult Still's disease.
- 2.8. Antiphospholipid antibodies syndrome.
- 2.9. Overlapping syndromes.
- 2.10. Undifferentiated connective tissue disease.
- 2.11. Idiopathic vasculitis predominantly affecting large vessels:
  - 2.11.1. Polymyalgia rheumatica and Temporal Giant Cell Arteritis;
  - 2.11.2. Takayasu disease;
  - 2.11.3. Isolated arteritis of the central nervous system.
- 2.12. Idiopathic vasculitis predominantly affecting medium-size vessels:
  - 2.12.1. Polyarteritis nodosa;
  - 2.12.2. Wegener's granulomatosis;
  - 2.12.3. Churg-Strauss syndrome;
  - 2.12.4. Kawasaki disease;
  - 2.12.5. Microscopic polyangiitis.
- 2.13. Idiopathic vasculitis affecting predominantly small vessels:
  - 2.13.1. Henoch-Schonlein purpura;
  - 2.13.2. Cutaneous leukocytoclastic vasculitis;
  - 2.13.4. Hypocomplementemic vasculitis;
  - 2.13.5. Crioglobulinemia.
- 2.14. Secondary vasculitis:
  - 2.14.1. Related with systemic rheumatic diseases;
  - 2.14.2. Related with neoplasia;

2.14.3. Related with infections;

2.14.4. Related with medicines;

2.14.5. Post-transplant.

2.15. Other vasculitis:

2.15.1. Behçet's syndrome;

2.15.2. Buerger's disease;

2.15.3. Relapsing polychondritis.

### 3. SPONDYLOARTHROPATHIES

3.1. Ankylosing spondylitis.

3.2. Psoriatic arthritis.

3.3. Reiter's syndrome and other reactive arthritis.

3.4. Arthropathies of chronic inflammatory bowel disease.

3.5. Undifferentiated spondyloarthropathies.

3.6. SAPHO.

### 4. RHEUMATIC FEVER

### 5. INFECTIOUS ARTHRITIS

5.1. Bacteria.

5.2. Virus.

5.3. Fungus.

5.4. Parasites.

## 6. CRYSTAL-ASSOCIATED ARTHROPATHIES

- 6.1. Gout.
- 6.2. Calcium pyrophosphate dihydrate deposition disease.
- 6.3. Others.

## 7. INTERMITTENT RHEUMATIC DISEASES

- 7.1. Palindromic rheumatism.
- 7.2. Intermittent hydrarthrosis.
- 7.3. Familial Mediterranean fever.

## 8. OSTEOPOROSIS

- 8.1. Primary.
- 8.2. Secondary.

## 9. BONE DISEASES

- 9.1. Osteoporosis.
- 9.2. Osteomalacia.
- 9.3. Paget's bone disease.
- 9.4. Algoneurodystrophy.
- 9.5. Osteonecrosis. 9.6. Renal osteodystrophy.
- 9.7. Others.

## 10. PERIARTICULAR RHEUMATIC OR SOFT TISSUE DISEASES

- 10.1. Tendonitis.

10.2. Tenosynovitis.

10.3. Enthesitis.

10.4. Bursitis.

10.5. Capsulitis.

10.6. Ligamentitis.

10.7. Others.

## 11. CHRONIC PAIN AND FATIGUE SYNDROMES

11.1. Fibromyalgia.

11.2. Myofascial syndrome.

11.3. Chronic fatigue syndrome.

## 12. TUMOURS

12.1. Joint.

12.2. Bone.

12.3. Bone metastases.

## 13. RHEUMATIC DISEASES INDUCED BY DRUGS, HAEMODIALYSIS AND SURGICAL PROCEDURES

## 14. MUSCULOSKELETAL MANIFESTATIONS OF NON-RHEUMATIC DISEASES

14.1. Haematological diseases:

14.1.1. Haemoglobinopathies;

14.1.2. Haemophilias;

14.1.3. Leukaemia;

14.1.4. Lymphomas;

14.1.5. Multiple myeloma.

14.2. Endocrine diseases:

14.2.1. Acromegaly;

14.2.2. Hyperthyroidism;

14.2.3. Hypothyroidism;

14.2.4. Hashimoto's thyroiditis;

14.2.5. Cushing's syndrome;

14.2.6. Hyperparathyroidism;

14.2.7. Hypoparathyroidism.

14.3. Metabolic diseases:

14.3.1. Diabetes.

14.3.2. Hemochromatosis.

14.3.3. Wilson's Disease.

14.3.4. Alkaptonuria / Ochronosis.

14.3.5. Hyperlipoproteinemias.

14.4. Hepatic diseases:

14.4.1. Infectious hepatitis.

14.4.2. Active chronic hepatitis.

14.4.3. Primary biliary cirrhosis.

14.5. Infiltrative diseases:

14.5.1. Sarcoidosis.

14.5.2. Gaucher, Fabry and Farber Diseases.

14.5.3. Lipochromic histiocytosis.

14.5.4. Multicentric reticulohistiocytosis.

14.5.5. Amyloidosis.

14.6. Skin diseases:

14.6.1. Acne.

14.6.2. Urticaria.

14.6.3. Panniculitis.

14.6.4. Neutrophilic dermatosis.

14.7. Immunodeficiency:

14.7.1. Common variable;

14.7.2. Combined;

14.7.3. Selective IgA;

14.7.4. Complement fractions;

14.7.5. Phagocytic cells;

14.8. Neuropathic arthropathy:

14.8.1. Tabes.

14.8.2. Syringomyelia.

14.8.3. Multiple sclerosis.

14.8.4. Pernicious anaemia.

14.8.5. Leprosy.

14.8.6. Alcoholism.

14.8.7. Others.

15. POST-TRAUMATIC ARTHRITIS

16. FOREIGN BODY SYNOVITIS

17. OTHERS

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