Acknowledging pain severity

Pain experienced by patients with autoinflammatory diseases is often not taken seriously due to lack of understanding as to how these innate genetic diseases impact the body. The threshold of pain is unique and different to each individual and can be varied based on their monogenic disorder type. It is important to remember that the pain experienced by patients during flares can be severe (not psychosomatic) and may require emergency care with pain control.

Autoinflammatory diseases may cause life-threatening issues, should patient be septic, have macrophage activation syndrome (MAS), present with hemophagocytic lymphohistiocytosis (HL), or cytokine storm. Other medical complications include bowel blockages, abdominal adhesions, heart attack, passing out risking a fall, etc. Severe pain can be the onset to these various issues and requires professional intervention.

Pain in children vs. pain in adults

Children and adults with treated autoinflammatory diseases may experience different levels of pain. Often NSAIDs tend to control the pain in toddlers and young children. However, when children reach puberty, the pain may intensify, possibly requiring additional medications. Adults may also respond well to NSAIDs. However, in some cases, alternative pain medications may be required.

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Prescribing pain medication

Pain medication may be prescribed by any doctor on the patient's care team, including a general practitioner, rheumatologist, immunologist, urgent/emergency room caregiver; however, this may vary from country to country. Long-term pain management in these diseases may require a specialist who understands how innate genetic diseases affect the body. Some patients may require high-dose pain management throughout the course of their life.

It is important to understand, that autoinflammatory diseases have no cure and lack of proper pain control can contribute to further physical damage if left untreated.

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PAIN MANAGEMENT

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ENGLISH

What are autoinflammatory diseases (AID)?

Autoinflammatory diseases, also known as periodic fever syndromes, are a group of rare disorders that, due to genetic "mutations," can cause an uncontrolled overactivity of the innate immune system. This can lead to episodes of spontaneous inflammation in one or more organs. Affected individuals may experience recurring flares associated with fever, serositis (inflammation of the serous membranes such as the chest, peritoneum, etc.), arthritis (joint inflammation), and PAIN.

Types of pain

Pain can impact the autoinflammatory patient in a variety of ways, in severity and may change over time. Types of pain are: acute, chronic, inflammatory, neuropathic, physiological, reflective, visceral, somatic, and may range from mild to severe, as reported by patients:

Aphthae (ulcers) and skin lesions: Mouth/genital ulcers, urticaria (hives)/rashes, psoriasis-like plaques, pustules, folliculitis (inflammation of the upper (outer) part of a hair follicle), ulcerations (tissue defects of the skin or mucous membrane), etc., can be painful, impact daily life, and require long-term treatment.

Chest/heart pain: Chest pain can include costochondritis, pericarditis, myocarditis, pleurisy, etc., and requires immediate medical attention.

Dermatological pain: itching, swelling, urticaria, psoriatic presentation, deep ulceration, bruising, hidradenitis suppurativa, folliculitis, etc.

Patient provided picture.

Gastrointestinal pain: abdominal pain, bloating, constipation, diarrhea, vomiting, nausea, and acid reflux. It can be mild or severe, dull or sharp, burning or achy, crampy or colicky, constant or intermittent, localized (in one spot) or generalized (all over) that often resolves by itself. However, other times, it can be a sign of a serious complication such as adhesions requiring urgent medical intervention. Severe abdominal pain can present like acute appendicitis, causing some patients to undergo unnecessary abdominal surgery, such as an appendectomy.

Headaches: These include headaches: migraine-like, tension, cluster, sinusitis, stress, temporomandibular joint (TMJ) issues, and ice pick. The pain can be stabbing, pulsating, and may also cause pressure behind the eyes. These headaches can lead to light and sound sensitivity, as well as vomiting, dizziness, tinnitus, drowsiness, nausea, and blurred vision.

Joint inflammation and pain may be associated with swelling: It can occur at rest or with physical activity in autoinflammatory patients. During a flare, these symptoms may lead to impaired mobility and require additional medication control.

Patient provided picture.

Leg pain:
Affects both children and adults. Leg pain may affect the joints and muscles, limiting ability to walk, and may be extremely severe. It is important to check vitamin D, as low blood levels may also increase the pain intensity. Supplement as recommended by your treating physician.

Lung pain: Pain in the lung area is a result of inflammation and can cause: wheezing, dry or productive (i.e. mucus-producing) cough/asthma (chronic inflammatory disease of the airways), difficulty breathing, discomfort or tightness in the chest, a sense of lung pain, air hunger, and feeling fatigued during physical exertion.

Spinal/back/hip pain: Ranges from muscle spasms to a shooting, burning or stabbing sensation in the spine. In addition, pain may radiate down the extremities or worsen with bending, twisting, lifting, standing, walking, or sleeping. Pain or stiffness may impact every part of the neck or back area, restricting range of motion and causing issues with posture, activities, and loss of motor function. Spondylitis (Inflammation of the vertebrae), disc compression/herniation, spinal fluid issues, etc., may all present. Hip pain can be a factor as well, including synovitis (inflammation of the joint membrane), bursitis, and tendonitis. Often back and hip pain may be misdiagnosed as osteoarthritis.

Treatment

Pain management control includes both over the counter (OTC) and prescription medications. Treatment with nonsteroidal anti-inflammatory drugs (NSAIDs) and acetaminophen can help to relieve pain, inflammation, and fever. Other drugs used include corticosteroids (see separate FMF & AID brochure), anticonvulsants, opioids, and topical analgesics (localized painkillers, including magic mouthwash for oral ulcers).

OTC pain medicines

Over-the-counter pain medications include: Acetaminophen/Paracetamol (Tylenol), Ibuprofen (Motrin, Advil), Naproxen (Aleve, Naprosyn), diclofenac (Voltaren), Novalgin/Metamizole/dipyrone.
These medications are useful because they help decrease pain, control swelling, and reduce inflammation.
Caution: Do not exceed the recommended dosage or combine these medications (i.e. aspirin with ibuprofen, or naproxen sodium) together unless indicated by a physician.

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Prescription pain meds

While OTC pain medications may be very effective, there are patients whose pain is not managed well or alleviated and require prescription drugs. Additionally, during a severe flare, strong medications may be required in a medical setting.

Prescription medications include the following categories:

Weaker-than-morphine opioids include: codeine, dihydrocodeine, meperidine, pentazocine, propoxyphene, and tramadol.

Morphine-equivalent opioids include: hydrocodone, morphine, and tapentadol.

Stronger-than-morphine opioids include: fentanyl, hydromorphone, methadone, oxycodone, and oxymorphone.

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Pain intensity

In a FMF & AID publication (2024), 44.9% of treated patients (surveyed) reported pain scores of +5 despite being on colchicine, biologics, or a combination of both. The pain intensity is variable and is measured with a Visual Analogue Scale (VAS).

All patients should understand the pain scale, enabling them to successfully manage their symptoms by determining their pain rating. Pain relief medications should be discussed and prescribed by your physician with instructions clarified for treating various breakthrough symptoms.
An emergency plan should also be made, should a hospital visit be required. It is appropriate for autoinflammatory patients to have "emergency medications" at home to alleviate sudden onset pain with doctor oversight.

Breakthrough symptoms

Many autoinflammatory patients on treatment for their disease have breakthrough symptoms or flares and require additional pain control medication.

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Undertreated patients

Patients whose disease is not well controlled or undiagnosed may experience pain due to being under medicated or untreated. Unfortunately, this is a common issue for autoinflammatory patients, and it is critical for the medical community to increase their knowledge about these diseases to assist patients with treatment and pain issues.

Alternative pain aids

The use of heat or cold therapy may help patients during their flares. These aids may include hot water bottle or heat pad, ice pack, hot bath/shower and hot/cold compresses.

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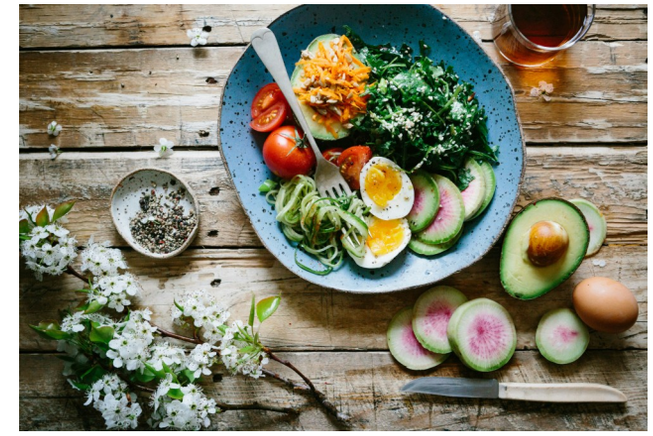
Natural/diet modifications

Natural aids such as herbal teas, ginger root, curcumin/capsaicin, CBD oil, tart cherry and others, may alleviate pain and improve overall immune health in some patients.

Diet modifications may be helpful in some cases, as patient may have unknown allergy or food intolerance (e.g. skin or GI), exacerbating their disease.



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