



# Chronic Nonbacterial Osteomyelitis and Chronic Recurrent Multifocal Osteomyelitis in Children

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## KEYWORDS

- Chronic nonbacterial osteomyelitis • Chronic recurrent multifocal osteomyelitis
- DIRA • Majeed syndrome • NSAID • Whole-body MRI

## KEY POINTS

- Chronic nonbacterial osteomyelitis (CNO; also known as chronic recurrent multifocal osteomyelitis) is an inflammatory/autoinflammatory bone disease that primarily affects children and adolescents. It is a diagnosis of exclusion.
- Often the diagnosis of CNO in children is delayed because of a lack of awareness and the occult nature of CNO. Prompt referral to pediatric rheumatology can help establish a diagnosis and determine appropriate treatment.
- Imaging studies, especially MRI with short tau inversion recovery, are essential diagnostic tools.
- Whole-body MRI is the gold standard for disease monitoring.
- Long-term treatment and follow-up are needed to prevent complications, such as vertebral compression fractures and leg-length discrepancies.

## INTRODUCTION

Chronic nonbacterial osteomyelitis (CNO; **a.k.a., chronic recurrent multifocal osteomyelitis**) is an inflammatory disorder that presents with bone pain arising from sterile osteomyelitis. It is primarily a pediatric disorder but can persist into adulthood or have an adult-onset presentation. The condition is difficult to diagnose, most commonly suspected to be infectious osteomyelitis or malignancy, with milder cases resembling growing pains. Children may have decreased physical function and poor school

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attendance when bone pain and inflammation are not controlled adequately. Delays in diagnosis can lead to permanent skeletal damage. The cause of the disease remains unknown for most but involves immune dysregulation resulting in inflammation of the bone and sometimes of other tissues, including skin, joints, and the intestine.

## NOMENCLATURE

The disease has gone by many names, making nomenclature complicated (**Box 1**). It was first described as a symmetric multifocal osteomyelitis and later given the name chronic recurrent multifocal osteomyelitis (CRMO).<sup>1</sup> However, because the disease may begin or stay unifocal, CRMO may not be an accurate term for these patients. Thus, the term *CNO* has been proposed as an umbrella term.

## INCIDENCE AND DEMOGRAPHICS

In 2011, the annual incidence of CNO in Germany was reported to be 0.4 per 100,000 children,<sup>2</sup> as compared with the reported incidence range of infectious osteomyelitis of 10 to 80 per 100,000 children per year.<sup>3</sup> However, during 2004 to 2014 in a single center in Germany, of the 109 children seen for osteomyelitis, 53% were categorized as infectious and 47% as noninfectious, unexpectedly similar proportions.<sup>3</sup> A single center in Britain reported increased patient referral for CNO after a letter was sent to all orthopedic centers to enhance recognition of the disease.<sup>4</sup> Although the actual incidence of CNO is likely to vary from one region to another, these studies suggest that it is more common than previously appreciated and underscore the importance of raising awareness of CNO.<sup>4</sup>

### Age

- The average age of disease onset is 9 to 10 years.<sup>2,4-8</sup> Rarely, disease onset occurs before 3 years of age.

### Sex

- Girls are more likely to be affected, with a female to male ratio of 2:1.<sup>2,4-8</sup>

#### Box 1

##### Reported terms of chronic nonbacterial osteomyelitis

Bone lesions of acne fulminans  
 Chronic multifocal cleidometaphyseal osteomyelitis  
 Chronic recurrent multifocal osteomyelitis  
 Chronic sclerosing osteitis  
 Chronic symmetric osteomyelitis  
 Clavicular hyperostosis and acne arthritis  
 Diffuse sclerosing osteomyelitis  
 Pustulotic arthro-osteitis  
 Sclerosing osteomyelitis of Garré  
 Sternocostoclavicular hyperostosis  
 Sternoclavicular pustulotic osteitis  
 Synovitis, acne, pustulosis, hyperostosis osteitis

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