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Editorial

Sickle Cell Disease: A Neglected Disease from the Osteo-Articular Point of View, in the Era of 'Evidence-Based Medicine

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In 1910, sickle cell disease (SCD) was clinically described for the first time by cardiologist Dr. James Herrick, in Chicago [1]. The first patient was Walter Clement Noel, a dentistry student African descent and native of the island of Grenada, Caribbean Sea [1]. In 1949, Linus Pauling., *et al*. discovered that the cause of SCD was a molecular injury in hemoglobin (Hb): the substitution of the amino acid valine for glutamic acid at position 6 of the beta chain [2]. In organic sites with low oxygen level (microcirculation), the polymerization of Hb, called HbS, occurs, which generates the typical deformity of red blood cells in this population: sickle-shaped as was called by Dr Herrick [3]. SCD is a group of genetic disorders, characterized by abnormal hemoglobin in red blood cells. This disease is more prevalent in people of African descent [4]. It has been estimated that in the USA there are 100,000 people suffering from SCD [5]. According to EMA [6], in Europe have been estimated that there are 2.6 SCD people per 10,000.

Although SCD is a primary hematological defect, sickle red blood cells produce, through complex mechanisms, damage to the vascular endothelium: SCD is a vasculopathy [7]. SCD affects the whole organism and the first manifestation is often the osteo-articular injury, in the form of painful SCD crisis: dactylitis.

Osteomyelitis and avascular necrosis or osteonecrosis are two of the main osteo-articular complications of SCD. Searching in PubMed with "sickle" AND "osteomyelitis" we can find 476 references: but no randomized clinical trial [8]. Would the clinical course of osteomyelitis in the SCD patient be the same as in a non-SCD patient? Why have no randomized clinical trial ever evaluated the clinical benefits and adverse events of the antibiotic regimens in sickle cell patients suffering from osteomyelitis? The sickle people are immunosuppressed due to the functional asplenia that suffers [9].

Similarly, if you search PubMed with "sickle" AND "avascular necrosis" you will find 454 references. You can look that there are only one randomized clinical trial (RCT) [10] and one Cochrane systematic review that found flawed methodology in that RCT [11].

In 2006, WHO declared SCD as a public health problem worldwide. It is not the frequency of SCD, but the high morbidity and disability generated by the osteo-articular damages SCD produces. It destroys the quality of life of the patient and family!

It is the lack of clinical trials in two major osteo-articular complications that causes SCD to be classified as a neglected disease. Medicine is an art, but with a scientific base.

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