Introduction: Red Ear Syndrome (RES) is an enigmatic disorder with approximately 100 published cases in literature. It is characterized by attacks of burning pain and erythema on the ear. RES is classified in idiopathic and secondary forms, often associated to primary headaches and upper cervical disorders respectively. The aim of this paper is to provide a review of studies which reports this poorly understood condition.

Methods: A Survey was made between November 2020 and January 2021 in the MEDLINE / PubMed (via national library of medicine) and Scopus (Elsevier) databases. The terms searched were "red ear syndrome", "red ear syndrome" AND "headache" and "red ear syndrome" AND "migraine". The bibliographic period range was from 1996 to 2021. Only files in English and Spanish were selected. Thus, the initial number of 432 articles were obtained.

After excluding duplicates, the number of articles was reduced to 230 different articles. After reading the titles and abstracts, 54 articles were selected for this review. As an exclusion criteria those papers about different disorders or syndromes, genetics and studies involving animals were excluded. Other exclusion criterium consisted of the term "red ear" being apart in the titles or abstracts, for example "inner ear". At the end of this initial analysis, other 4 articles were selected for full reading due to their importance for the topic.

We reviewed all previously described cases and 58 articles were selected following the Preferred Reporting Items for Systematic Review and Meta-Analysis (PRISMA) protocol (Figure 1). All the 94 patients collected from case reports were placed into idiopathic or secondary RES groups.

Results: In both groups there was a female to male predilection ratio; unilateral attacks were more common, and the attacks could last from seconds to hours and occur daily. In the idiopathic RES, 73.7% of patients the attacks were associated to primary headaches (Migraine and Trigeminal Autonomic Cephalalgias) and 20% had both condition isolated, and the most common trigger was tactile stimuli. On the other hand, in secondary RES, the most common trigger was head movement.

Data revealed that during episodes of RES the presence of extraauricular symptoms is prevalent, especially pain. It was always present in the ear, and could spread to adjacent regions (Figure 2). The more intense the color, greater the number of patients who reported symptoms in that region. In 47.5% of idiopathic RES patients the pain had extended beyond the ear, mostly to the trigeminal innervation territories. In the secondary RES patients, overspreading of pain was clearly more frequent: 83.8% of patients. It was observed that in secondary cases there is a greater range of pain beyond the ear for cervical root innervation territories C2 to C5. Patients can also experience autonomic and vestibulocochlear symptoms.

Conclusion: Our systematic review showed important clinical differences between primary and secondary RES. These results could shed light on the knowledge of this peculiar condition. Moreover, a solid diagnostic criteria are important to identify RES patients and provide grounds for more in-depth investigation of the Red ear Syndrome.