

Parry-Romberg Syndrome

a rare disorder

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# Fall 2015

Parry Romberg Syndrome

Parry Romberg Syndrome was first described by Caleb Hillier Parry in 1825[1] then again by Mortiz Heinrich Romberg in 1846[2]. It is defined as a rare degenerative disorder that is distinguished by the slow gradual shrinkage and wasting away of skin, muscles, bones, soft tissues, fat and cartilage. The degradation of the skin and subcutaneous soft tissues are usually unilateral of the face and can be self-limiting. The exact etiology and pathophysiology of this syndrome is unknown. Theories such as trauma, viral infections including *Borrelia burgdorferi*, hormone changes, genetics, autoimmune disorders, peripheral trigeminal neuritis increase of cervical sympathetic nerve activity and changes of fat metabolism, are possible suggested factors playing a role in pathogenesis of this condition [3]. A recent study done in 2014 suggest that a relation between Perry Romberg and a disorder of the neural cell migration is present and could be a reason for the malformation. [4] It is likely that the disease results from different onsets and is different person to person. Unfortunately, there is no cure or treatment that can prevent Parry Romberg syndrome. Some individuals have been reported to use prescription drugs such as Methotrexate and Prednisolone in an attempt to stop or slow the progression with varying results. Reconstructive or microvascular surgery may be needed to repair tissue. Recent studies have also used stem cells in conjunction with fat grafts to restore the soft tissue, marking it superior to conventional lipo-injection. [5] The timing of surgical intervention is controversial; some prefer to wait until the disease has stabilized while others recommend early intervention.[6]

Parry Romberg Syndrome typically appears in late childhood or teen years, between ages five to fifteen years old however, the disorder has been described in infants and individuals of fifty years and older. Parry Romberg Syndrome has a higher incidence in females over males and mostly affects the left side over the right.[7] The years of deterioration can last at varying lengths from two to ten years concluding at a stable state, however some individuals have a longer lasting active period.[6] According to the he National Organization for Rare Disorders (NORD), any disease affecting fewer than 200,000 Americans is considered rare. Because this rare disorder often goes undiagnosed or misdiagnosed, finding the quantity of Parry-Romberg syndrome in the general human population is challenging. Physicians studying the disorder have estimated that Parry-Romberg may affect as many as 1 in 250,000 people in the general population, as of 2013.[8]

Parry-Romberg Syndrome may also be accompanied with neurological abnormalities like migraines, seizures, strokes, lesions, atrophy and hemi-brain atrophy; optic nerve damage like uveitis, decreased pressure, retraction, vision loss, eyelid asymmetry; bone loss dermatological concerns like alopecia, pigmentation irregularities, epidermal atrophy, scleroderma en coupe sabre; and dental changes like asymmetry of the maxilla lips tongue and gum (usually deviates to the affected side), difficulty opening the mouth(however speech is unaffected), malocclusion, and root malformation, eruption or resorption.[9][10] In severe cases where the disease progresses beyond the face to the body, the individual will experience atrophy of the arms, torso and leg.[11] However every case is different so an individual can express most of these abnormalities, a few or none.

The disease is self-limiting and has no definite cure.[12] The patients affected usually seek help from several professionals including dermatologists, dentists and psychologists.[13] Surgical treatment is usually based on reposition of adipose tissue that was lost. Autogenous fat grafts, cartilage grafts, silicone injections and prostheses, bovine collagen, inorganic implants and cell fat mixed with platelet gel are some alternatives to aesthetic correction of the atrophy.[14]

Parry-Romberg syndrome is a rare disorder where the cause is unknown. Theories are suggested however no exact cause has been proven as the sole reason for this syndrome. It can be distinguished by the unilateral degradation of the face and can extend to the body in more severe cases. Unfortunately there is no cure or proven way to slow the deterioration of the skin and bones, however it is self-limiting and varies person to person.

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