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Aspirin responsive erythromelalgia in JAK2-thrombocythemia and incurable inherited erythrothermalgia in neuropathic Nav1.7 sodium channelopathy: From Mitchell 1878 to Michiels 2017

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Introduction: The article reports on the nosologic classification and common etiologic pathways of aspirin responsive erythromelalgia in clonal JAK2 thrombocythema and incurable inherited autosomal dominant erythrothermalgia in Nav1.7 channelopathy.

Areas Covered: Aspirin responsive platelet-mediated erythromelalgic arteriolar inflammation and thrombotic manifestations in thrombocythemia are caused by prostaglandin endoperoxides released from spontaneous activation, release reaction and aggregagation of hypersensitive constitutively activated platelets due to gain-of-function mutations in the JAK2, TPO, MPL and CALR genes (sticky platelet syndrome). Incurable inherited erythrothermalgia is a rare autosomal dominant neuropathy due to gain function mutations in the *SCN9A* gene that result in warmth-induced burning pain of the body and both legs sparing the toes and fingers caused by hyperexcitable action potentials firing of the Nav1.7 mutated small C-fiber DRG nociceptive sensory neurons of the skin.

Expert Opinion: Acetyl salicylic acid (aspirin) is a wonder drug in the treatment of symptomatic thrombocythemia patients with erythromelalgia but not sodium salicylate, coumadin and the platelet inhibiting agents ticlopedin and dipyridamole. The JAK2, MPL, CALR or TPO gain of function mutations constitutively activate megakaryopoesis and increase the production of hypersensitive platelets as the cause of aspirin responsive erythromelalgia. Incurable inherited erythrothermalgia is a congenital autosomal dominant neuropathic Nav1.7 channelopathy of the afferent C-fibers neurons and blood flow regulating system in the arteriole venule shunt of the skin caused by a gain-of-function mutation in the SCN9A gene.

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