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Common etiologic pathways in platelet-mediated aspirin responsive erythromelalgia and arteriolar platelet thrombophilia in JAK2, MPL or CALR mutated thrombocythemia and incurable Nav1.7 neuropathic erythrothermalgia due to gain of function mutations in the *SCN9A* gene

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The nosologic classification of burning, painful, red congested extremities in the 1990s included aspirin responsive erythromelalgia and microvascular disturbances in thrombocythemia versus aspirin resitent primary erythermalgia, an incurable autosomal dominant Nav1.7 neuropathic erythrothermalgia due to gain of function mutations in the *SCN9A* gene. Both conditions should not be confounded with aspirin resistant erythermalgia secondary to cutaneous vasculitis, systemic lupus erythematosus, rheumatoid arthritis, associated with hypertension or elicited by vasoactive drugs. Michiels discovered between 1975 and 2017 common etiologic pathways in platelet-mediated aspirin responsive erythromelalgia in *JAK2*-thrombocythemia in patients with essential thrombocythemia (ET) and polycythemia vera (PV) and incurable neuropathic inherited erythrothermalgia in neuropathic Nav1.7 of dorsal root ganglia. The two disorders are defects in the thermoregulatory and pain-sensing nociceptive afferent sensory C-fibers neurons and blood flow regulating the system in the arteriole-capillary arterial-venues (AV) shunt. Aspirin responsive von Willebrand factor-platelet-mediated erythromelalgia arteriolar inflammation and thrombosis followed by microvascular ocular, cerebral and coronary thrombosis in thrombocythemia (EMT) patients are caused by prostaglandin endoperoxides released from spontaneous activation, release reaction and aggregation of hypersensitive constitutively activated platelets due to a gain of function mutations in the *JAK2, TPO, MPL* and *CALR* genes (Sticky Platelet Syndrome). Incurable inherited erythrothermalgia (IE) and Paroxysmal Erythromelalgia Extreme Pain Disorders (PEPD) are incurable dominant congenital neuropathic pain conditions caused by gain of function mutations in the *SCN9A* gene as the cause of Nav1.7 sodium channel protein in the dorsal root ganglion (DRG) nociceptive neurons of afferent C-fibers in the subcutaneous arteriole-capillary arterial-venues (AV) shunt. Targeted curative management of *JAK2*-Thrombocythemia in ET and PV patients with aspirin responsive erythromelalgia and its microvascular and major thrombotic complications caused by a gain of function mutation in J*AK2, MPL, CAL* or *TPO* gene is feasible when adequately diagnosed in time. Inherited erythromelalgia (IEM) is a congenital dominant neuropathic Nav1.7 channelopathy caused by a gain of function mutation in the *SCN9A* gene and remains incurable and a great suffer for affected patients recently labeled as the 'Man on Fire Syndrome' in medicine anno 2016, that cries for targeted initiatives to develop novel effective analgesics by pharmaceutical companies.

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