



NEVUS FLAMMEUS

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Nevus flammeus, also known as port wine stain, is a congenital capillary malformation. It may present at birth as a flat, painless, blanchable, pink to red lesion that may occur anywhere on the body with unilateral or segmental distribution. When present on the face, the lesions usually follow trigeminal nerve branches distribution and may extend into mucosal surfaces. The lesions do not regress and may increase in size as the child grows. Additionally, they may become thicker, darker, and nodular over time.

The lesions may present as isolated cutaneous anomalies or be part of a complex syndrome. As isolated lesions, they are generally sporadic; however, familial cases have been reported. As part of a syndrome, they are most commonly associated with **Sturge-Weber**, Klippel-Trenaunay, Parkes-Weber, Servelle-Martorell, Proteus, Bannayan-Riley-Ruvalcaba, and CLOVES syndromes.

The diagnosis of nevus flammeus can be made based upon the history and physical examination. However, imaging studies might be necessary to rule out associated syndromes. Patients presenting with lesions on the eyelids should be referred for ophthalmological evaluation to rule out glaucoma. Early treatment of nevus flammeus with laser and light therapy may prevent thickening and development of nodularity. Pulsed dye laser is considered the standard of care for treatment, while cryotherapy, electrocautery, and excision should be avoided to prevent scarring.

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