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NEUROLOGY, NEUROIMAGING, AND OPHTHALMOLOGY CONSENSUS RECOMMENDATIONS FOR MANAGEMENT OF STURGE-WEBER SYNDROME

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Background: Sturge-Weber syndrome (SWS) involves the skin, brain, and eyes. The dermatologic manifestation is the facial port wine birthmark (PWB), while there are several potential neurologic and ophthalmologic manifestations, notably epilepsy and glaucoma, respectively. This consensus aims to develop evidence-based expert-defined recommendations for management of the neurologic and ophthalmic features and provides a framework for dermatologists to determine workup for patients with PWB who seek laser treatment.

Study Design/Materials and Method: Thirteen national experts in neurology, radiology, and ophthalmology were assembled as part of a larger consensus statement for the management of SWS. Key topics and questions regarding risk stratification, referral indications, and treatment were formulated. A systematic PubMed search was performed. Evidence-based recommendations were developed.

Results: High-risk PWB distributions involve the

hemifacial, forehead, or median locations, including the upper eyelids, especially when concerned for glaucoma. Any child with a high-risk facial PWB should be referred to a pediatric neurologist and pediatric ophthalmologist for a baseline evaluation, with periodic follow-up. Routine screening for brain involvement is not recommended for newborns and infants with a high-risk PWB and no history of seizures or neurological symptoms but can be performed in cases of extreme parental anxiety, an abnormal EEG, or when presymptomatic treatment is contemplated such as with extensive bilateral PWB. In children with stable neurocognitive symptoms, routine follow-up neuroimaging is not advised. In adults with a high-risk PWB and no prior imaging, neuroimaging should be obtained, but follow-up neuroimaging is not recommended in adults with established SWS and stable neurocognitive symptoms. The treatment of glaucoma varies depending on the patient's age and clinical presentation.

Conclusion: Recommendations were developed by experts in neurology, neuroradiology, and ophthalmology. These guidelines can guide evidence-based discussions between patients and providers and increase dermatologists' awareness regarding when and what workup is necessary in patients seeking laser treatment for PWB.