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Evolving Clinical Presentation and Assessment of Pheochromocytoma

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Pheochromocytoma (PHEO) is a neuroendocrine lesion in the adrenal medulla composed of chromaffin cells producing excess amount of catecholamines. These tumoral cells have the property to synthesize, metabolize, store, and secrete catecholamines and their metabolites. The clinical symptomatology is derived from the peripheral tissue effect of norepinephrine, epinephrine, and their by-products. Morbidity and mortality is increased due to the delay in the diagnosis and treatment. A high index of suspicion leads to testing for PHEO through biochemical, imaging, and genetic studies. Dilemma in its assessment comes about when the clinical picture is beset by too much catecholamine secretory periodicity, too little catecholamine secretion, in lesions less than 1 cm, in exclusively dopamine-secreting tumors, and in the unavailability of biochemical tests and imaging procedures.

In this review, the discussion is centered on the progress in the approach of early diagnosis of pheochromocytoma through improved clinical and biochemical assessment. Emphasis is made on the early recognition of evolving clinical presentations, with the introduction of cardiovascular imaging, 2D echocardiogram, and cardiac MRI in the early diagnosis of patients with no risk factors and with equivocal biochemical and imaging results yet present with cardiovascular events. An algorithm is established for easy guide for clinicians.