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Contemporary Perioperative and Anesthetic Management of Pheochromocytoma and Paraganglioma

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Keywords

• Pheochromocytoma • Paraganglioma • α-blockade • Catecholamine
• Phenoxybenzamine • Prazosin • Doxazosin

Key points

- The care of patients with pheochromocytoma and paraganglioma (PPGL) presenting for surgical resection presents a challenge for the anesthesiologist.
- There is no evidence to specifically recommend the choice of preoperative pharmacologic preparation.
- Practical considerations for choice of preoperative pharmacological management include drug availability and cost.
- A detailed understanding of catecholamine biosynthesis, metabolism, and physiology provides a sound basis for understanding the cardiovascular manifestations and risks, diagnostic evaluation, preoperative preparation, and perioperative hemodynamic management of patients with PPGL presenting for surgical resection.
- Knowledge of the preoperative risk factors and perioperative events that increase the risk for hemodynamic instability provides a sound physiologic and pharmacologic basis for optimizing perioperative outcome.

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INTRODUCTION

Pheochromocytomas are catecholamine-secreting tumors that arise from the chromaffin cells located within the adrenal medulla. Paragangliomas are also catecholamine-secreting tumors arising from extra-adrenal chromaffin cells located along the sympathetic paravertebral ganglia of the pelvis, abdomen, and thorax. They are clinically relevant in perioperative medicine because they can produce and secrete large amounts of one or more catecholamines: epinephrine (E), norepinephrine (NE), and, uncommonly, dopamine. Because they produce clinically similar symptoms based on the type, amount, and secretory pattern of excess catecholamine secretion, they may be collectively referred to as pheochromocytoma and paraganglioma (PPGL).

The majority (80%–85%) of PPGL are located within the adrenal gland. PPGL are rare, with annual estimated yearly incidence of 2 to 8 cases per million [1], whereas the prevalence as a cause of secondary hypertension in the outpatient population has been estimated to be 0.1% to 0.6% [2,3]. The historically quoted "*Rule of 10s*" (10% of PPGL are extra-adrenal, 10% are malignant, 10% are bilateral, and 10% are familial) is incorrect particularly in that at least 32% of PPGL are familial [4]. In addition, the prevalence of metastatic disease varies between 10% and 17%, and in certain inherited subtypes of PPGL, the prevalence may be as high 40% or more of patients [5].

CATECHOLAMINE SYNTHESIS, METABOLISM, AND PHYSIOLOGY

A basic knowledge of the synthesis, secretion, metabolism, and physiologic actions of catecholamines is essential to understanding the clinical presentation, diagnostic evaluation, and perioperative pharmacologic management of cardiovascular manifestations of PPGL (Fig. 1) [6,7]. Catecholamines are synthesized within and subsequently secreted from chromaffin cells. The initial and rate-limiting step in catecholamine synthesis is the enzymatic hydroxylation of L-tyrosine to L-3,4-dihydroxyphenylalanine (DOPA) by tyrosine hydroxylase. Subsequently, DOPA undergoes decarboxylation by aromatic L-amino acid decarboxylase (AADC) to dopamine. Dopamine is then actively transported into

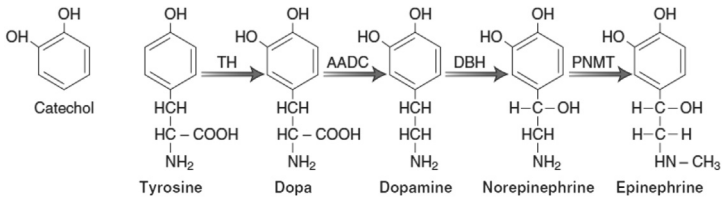

Fig. 1. Biosynthetic pathway for catecholamine metabolism. (*Modified from* Dluhy RG, Lawrence JE, Williams GH. Endocrine hypertension. In: Larsen PR, Kronenberg HM, Melmed S, et al. editors. Williams textbook of endocrinology. 10th edition. Philadelphia: Saunders; 2003. p. 555; with permission.)