PHEOCHROMOCYTOMA

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Question

A 10-year-old male presents to the OR for resection of a pheochromocytoma. Preoperatively, the patient was managed on phenoxybenzamine and propranolol. Which of the following preoperative signs and/or symptoms most likely indicate that this patient is optimized for surgery?

A. Documented blood pressures <50th percentile for age and height
B. Frequent PVCs on telemetry
C. Symptomatic orthostatic hypotension
D. Intermittent headaches and diaphoresis

Key Points

- Pheochromocytomas are catecholamine-secreting tumors that cause excessive α- and β-adrenergic stimulation.[1]
- Preoperative α-blockade is key in reducing intraoperative hemodynamic swings and surgical morbidity and mortality. β-blockade should only be initiated after adequate α- blockade to avoid dangerous increases in SVR due to unopposed α-mediated vasoconstriction.[2]
- Intraoperative management focuses on avoiding drug-induced catecholamine release and minimizing hemodynamic swings associated with intubation, surgical manipulation, and tumor removal.[2]

Description

- Pheochromocytomas are tumors of neural crest origin that produce and secrete norepinephrine, epinephrine, and dopamine. Although they are predominantly found in the adrenal medulla in adults, a significantly higher percentage of tumors in children are extra-adrenal. They have a strong genetic predisposition and almost 40% of cases in children are associated with genetic syndromes, including multiple endocrine neoplasia (MEN) 2A and 2B, Neurofibromatosis type 1, and von Hippel-Lindau syndrome.[2]
- A two-stage process of confirming and localizing the tumor via biochemical testing and imaging, respectively, is necessary. In children, measuring **urinary metanephrines** is the most useful test in confirming the diagnosis of pheochromocytoma. CT or MRI can accurately locate most tumor(s), while MBIG scintigraphy is used to locate small foci.[3]

Pathophysiology

Clinical presentation includes sudden severe headaches, hypertension, perspiration, visual problems, weight loss, pallor, nausea, polyuria, and polydipsia. A **classic triad of paroxysmal diaphoresis, palpitations, and headaches** is often described, **while nausea is an especially common complaint in the pediatric population**.[2]

Effects of adrenergic receptor stimulation[4]:
α_1-receptor: $\Uparrow$ PVR/arterial BP, $\Downarrow$ circulating blood volume, congestive heart failure, renal failure, cerebral hemorrhage
β_1-receptor: $\Uparrow$ myocyte automaticity, $\Uparrow$ ventricular ectopy

Perioperative Management

The mainstay of preoperative preparation prior to surgery is blood pressure control with α-blockade. Typically, **phenoxybenzamine**, a long-acting α-antagonist administered orally, is **titrated to blood pressures <50th percentile for age and height in children**.[1] β-blockade can then be added to control tachycardia and dysrhythmias. If β-blockade is initiated first, norepinephrine and epinephrine will produce unopposed α-vasoconstriction that cannot be offset by β_2-mediated vasodilation. Additionally, to restore normal circulating blood volume, a high sodium diet and increased fluid intake (1.5X maintenance) are recommended.[2]

Preoperative Preparation Goals	Laboratory Investigations
Blood pressure control Reversal of circulating volume depletion Heart rate and rhythm control, Correction of blood glucose and electrolyte disturbances	CBC Electrolyte panel, creatinine, blood glucose CT/MRI EKG, echocardiogram

Expected periods of intraoperative hemodynamic lability:

- Intubation, incision, insufflation, tumor manipulation → profound hypertension, tachyarrhythmias, hyperglycemia
- Tumor devascularization: hypotension, hypoglycemia[5]

Intraoperative Management of Pheochromocytoma[2]		
Monitoring and Vascular Access	**Available Medications**	**Medications to Avoid** (Potential for catecholamine increase)
Standard ASA monitors	Anxiolytic premedication	Ketamine
5-lead EKG	Magnesium sulfate (inhibits release of catecholamines)	Desflurane
Arterial line (before or after induction depending on patient cooperation)	Short-acting opioids (fentanyl, remifentanil)	Morphine
Consider central venous catheter	Dexmedetomidine (centrally acting α_2 agonist)	Pancuronium/atracurium
Large bore peripheral venous access	Vasodilators (nitroprusside, phentolamine, nicardipine)	Succinylcholine
Urinary catheter	β-blockers (esmolol, labetalol)	Ephedrine
Consider TEE in patients with cardiomyopathy	Vasopressors (norepinephrine, vasopressin)	Droperidol
	Insulin and dextrose-containing fluids	Metoclopramide

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Answer: A. Preoperative α-blockade and volume repletion is key in reducing intraoperative hemodynamic swings. Frequent PVCs and intermittent headaches and diaphoresis allude to ongoing catecholamine surges, while symptomatic orthostatic hypotension demonstrates inadequate volume repletion. In children, α-blockade should be titrated until blood pressures are <50th percentile for age and height.

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