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INDICATIONS FOR TESTING

- Signs and symptoms of PPGL (eg, episodic hypertension, headache, tachycardia)
- Cardiovascular event consistent with PPGL in the presence of signs and symptoms
- Suggestive incidental findings on imaging
- Diagnosis or family history of an associated hereditary syndrome (eg, *SDH*-associated disorders, MEN2, NF1, VHL)
- Type 2 diabetes with a body mass index <25 kg/m²

ORDER

Plasma free metanephrines test
(in high-risk patients)
AND/OR
24-hr urine fractionated metanephrines test
(in low-risk patients)

Not elevated

Mildly elevated or indeterminate

Moderate to highly elevated

PPGL unlikely; if high suspicion exists, repeat testing in 3-6 mos

Periodic surveillance may be appropriate in high-risk patients, depending on clinical circumstances

CONSIDER
Potential false-positive results
(due to influence of diet, medications, inappropriate sampling conditions)

Additional evaluation if suspicion remains

High suspicion for PPGL

Repeat metanephrines testing under controlled conditions

Not elevated

Elevated

PPGL unlikely

PPGL likely

If high suspicion exists, repeat testing in 6-12 mos

Proceed with imaging

CONSIDER
Genetic testing

Abbreviations

MEN2 Multiple endocrine neoplasia type 2
NF1 Neurofibromatosis type 1
PPGL Pheochromocytoma and paraganglioma
VHL Von Hippel-Lindau syndrome