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Precision Surgical Management of Pheochromocytoma and Paraganglioma: A Tertiary Referral Center Experience

Ivan Vasilevski, MD, PhD*

Department of General, Abdominal and Vascular Surgery, Medical Institute of the Ministry of Interior, Sofia, Bulgaria.

ORCID: 0009-0008-3242-8227.

***Correspondence:**
Ivan Vasilevski, MD, PhD, Department of General, Abdominal and Vascular Surgery, Medical Institute of the Ministry of Interior, Sofia, Bulgaria.

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ABSTRACT

Background: *Pheochromocytoma and paraganglioma (PPGL) are rare catecholamine-secreting neuroendocrine tumors associated with substantial cardiovascular morbidity and potentially life-threatening complications if left untreated. Surgical resection remains the only curative treatment. Advances in functional imaging, genetic stratification, and minimally invasive surgical techniques have significantly refined contemporary management strategies.*

Objective: *To summarize current principles of precision surgical management in PPGL and to present institutional experience from a tertiary referral center.*

Methods: *This study combines a focused literature review with a retrospective analysis of seven consecutive patients who underwent adrenalectomy for pheochromocytoma at a tertiary referral center between 2023 and 2025. Preoperative assessment, genetic evaluation, surgical approach, perioperative outcomes, and follow-up data (10–12 months) were analyzed descriptively.*

Results: *Modern cross-sectional and functional imaging enabled accurate tumor localization and individualized operative planning. Germline genetic testing informed the extent of adrenal resection, particularly in hereditary syndromes such as multiple endocrine neoplasia type 2 and von Hippel–Lindau disease. All patients underwent minimally invasive unilateral adrenalectomy. One patient experienced an intraoperative hypertensive crisis, which was successfully managed. The median tumor size was 5.2 cm. No postoperative complications or perioperative mortality were observed. Favorable clinical outcomes were maintained during a median follow-up of 11 months, with no evidence of local recurrence or metastatic progression.*

Conclusion: *Precision-based surgical management integrating advanced imaging, biochemical profiling, and genetic stratification enables safe and effective treatment of PPGL. Even in small-volume series, a structured multidisciplinary approach can achieve excellent perioperative outcomes while preserving adrenal function when appropriate.*

Keywords

Pheochromocytoma, Paraganglioma, Adrenalectomy, Genetic testing, RET mutation, Cortical-sparing surgery, Precision surgery, Endocrine surgery.

Abbreviations

PPGL: Pheochromocytoma and paraganglioma, PCC: Pheochromocytoma, PGL: Paraganglioma, NET: Neuroendocrine tumor, NGS: Next-generation sequencing, CT: Computed tomography, MRI: Magnetic resonance imaging, ^123I-MIBG: Iodine-123 metaiodobenzylguanidine, ^123I-MIBG SPECT/CT: Iodine-123 metaiodobenzylguanidine single-photon emission computed tomography/computed tomography, PET/CT: Positron emission tomography/computed tomography, TA: Transabdominal

approach, PRA: Posterior retroperitoneoscopic approach, BMI: Body mass index, VHL: Von Hippel–Lindau disease, SDHB / SDHC / SDHD: Succinate dehydrogenase subunit B / C / D, MEN2A / MEN2B: Multiple endocrine neoplasia type 2A / type 2B, WHO: World Health Organization.

Introduction

Pheochromocytomas (PCCs) and paragangliomas (PGLs), collectively referred to as pheochromocytoma–paraganglioma (PPGL), are rare catecholamine-secreting neuroendocrine tumors arising from adrenal medullary chromaffin cells or extra-adrenal paraganglia [1,2]. Despite their low incidence, PPGLs are associated with substantial cardiovascular morbidity and potential mortality due to catecholamine excess and perioperative hemodynamic instability [1,3].

The 2022 World Health Organization (WHO) classification recognizes all PPGLs as tumors with metastatic potential, eliminating the historical distinction between "benign" and "malignant" pheochromocytoma [2]. This paradigm shift has reshaped long-term surveillance strategies and emphasizes risk-adapted management.

Over the past two decades, advances in molecular genetics, functional imaging, and minimally invasive adrenal surgery have significantly refined therapeutic strategies [3-5]. Approximately 30–40% of patients carry germline pathogenic variants, making PPGL one of the most heritable solid tumors in adults [3,6]. Increasing recognition of genotype–phenotype correlations has influenced operative planning, particularly regarding the extent of resection, the role of cortical-sparing adrenalectomy, and individualized surveillance protocols [4,7].

Although numerous surgical series have been published, heterogeneity persists regarding integration of genetic stratification into operative decision-making and reporting of contemporary outcomes. Real-world data reflecting structured precision-based management in tertiary referral centers remain limited.

The present study evaluates surgical strategy, perioperative outcomes, and the clinical integration of genetic stratification in patients with PPGL treated at a tertiary endocrine surgery center.

Genetic Stratification and Surgical Implications

Molecular Landscape

Up to 40% of patients with PPGL harbor germline mutations in susceptibility genes, most commonly SDHx (SDHA, SDHB, SDHC, SDHD), VHL, RET, NF1, MAX, and TMEM127 [3,6,8]. More than 20 susceptibility genes have been identified, reflecting the molecular heterogeneity of these tumors.

Transcriptomic analyses classify PPGL into three principal molecular clusters:

- Pseudohypoxia-related (e.g., SDHx, VHL)
- Kinase signaling-related (e.g., RET, NF1, TMEM127, MAX)
- Wnt pathway–altered tumors

These molecular subtypes correlate with metastatic risk, tumor location, imaging phenotype, and long-term outcomes [4,8].

Genotype–Phenotype Correlation

Genotype–phenotype correlations have direct surgical implications. SDHB mutations are associated with extra-adrenal location and increased metastatic risk [6,9]. In contrast, VHL- and RET-associated tumors frequently present as bilateral adrenal pheochromocytomas, often at younger age [3,10]. MAX and TMEM127 mutations are also linked to bilateral or multifocal adrenal disease [6,8].

These distinctions influence:

- Extent of adrenal resection
- Consideration of cortical-sparing adrenalectomy
- Surveillance intensity and duration
- Preoperative staging strategies

Patients with high-risk genotypes, particularly SDHB, require comprehensive staging and prolonged follow-up [6,9].

Clinical Integration of Genetic Testing

Current international guidelines recommend universal germline testing for all patients with PPGL, irrespective of age or family history [3,7]. Next-generation sequencing (NGS) panels have become the standard diagnostic approach due to high sensitivity and diagnostic yield [6].

Genetic profiling now directly informs:

- Surgical extent (total vs cortical-sparing adrenalectomy)
- Risk-adapted follow-up
- Screening of first-degree relatives
- Long-term metastatic surveillance

Accordingly, contemporary PPGL management is increasingly genotype-driven and requires coordinated multidisciplinary care.

Imaging Strategy in the Era of Precision Surgery

Accurate tumor localization, staging, and assessment of locoregional extension are fundamental prerequisites for safe and individualized preoperative surgical management of PPGL [1,3]. Contemporary imaging strategies integrate anatomical and functional modalities within a genotype-informed diagnostic algorithm.

Anatomical Imaging

Following biochemical confirmation of PPGL, cross-sectional anatomical imaging—most commonly contrast-enhanced computed tomography (CT) and/or magnetic resonance imaging (MRI)—is mandatory for preoperative localization and operative planning [1,7].

Contrast-enhanced CT remains the first-line modality in most patients due to its high spatial resolution and widespread availability. On non-contrast CT, pheochromocytomas typically demonstrate relatively high attenuation (>10 HU), reflecting

their hypervascular nature. During arterial-phase imaging, they typically demonstrate intense enhancement with variable washout patterns. Lesions are frequently heterogeneous, with areas of cystic degeneration, necrosis, hemorrhage, or occasional calcifications [7,11].

MRI provides superior soft-tissue contrast and is particularly valuable in:

- younger patients,
- individuals with contraindications to iodinated contrast,
- suspected extra-adrenal or head and neck paragangliomas,
- hereditary syndromes requiring repeated imaging surveillance [1,7].

On T2-weighted sequences, pheochromocytomas often exhibit marked hyperintensity ("light-bulb sign"), although this feature is not consistently present. Dynamic contrast-enhanced sequences demonstrate avid enhancement corresponding to tumor vascularity [11].

Beyond lesion detection, anatomical imaging defines:

- tumor size,
- tumor–vascular interface (IVC, renal vein, aorta),
- local invasion,
- feasibility of minimally invasive adrenalectomy.

These parameters directly influence surgical approach selection (transperitoneal vs. retroperitoneoscopic vs. open), operative positioning, and vascular control strategy [5].

Institutional Imaging Experience

In our cohort, anatomical imaging directly influenced operative strategy, vascular control planning, and perioperative risk stratification.

Figure 1 demonstrates a contrast-enhanced CT scan revealing a heterogeneous right adrenal mass measuring 4.8 × 3.8 cm, exerting compression on the inferior vena cava and abutting the upper pole of the right kidney. The intimate vascular relationship influenced operative positioning and early vascular control strategy.

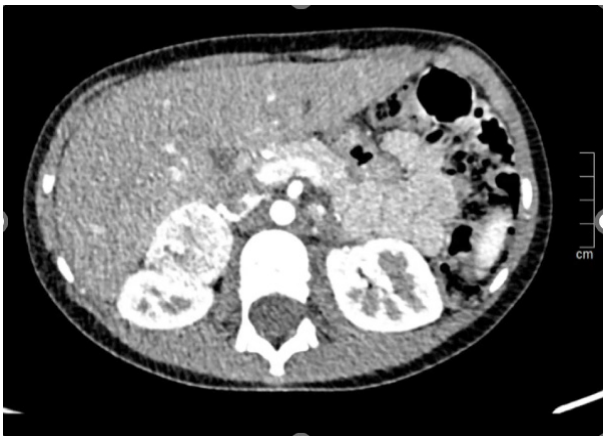

Figure 1: Contrast-enhanced CT scan demonstrating a heterogeneous adrenal mass measuring 4.8 × 3.8 cm, exerting compression on the inferior vena cava and abutting the upper pole of the right kidney.

Figure 2 illustrates a contrast-enhanced CT showing a heterogeneous solid mass measuring approximately 6 cm in the region of the left adrenal gland. Tumor size and radiological heterogeneity suggested increased risk of intraoperative catecholamine surges, prompting meticulous preoperative hemodynamic preparation.

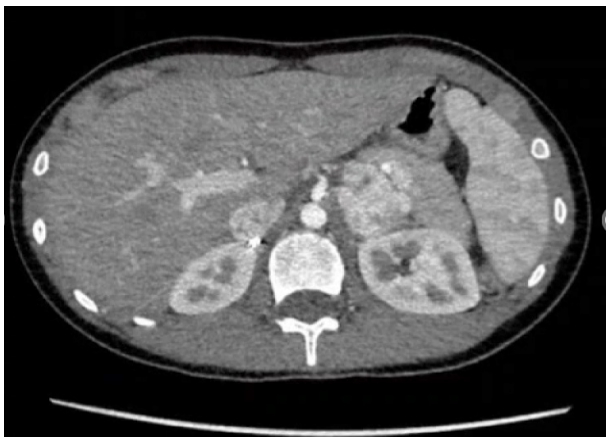

Figure 2: Contrast-enhanced computed tomography demonstrating a heterogeneous solid mass measuring approximately 6 cm, located in the region of the left adrenal gland.

Radiological characteristics were systematically integrated into intraoperative risk assessment and surgical approach selection.

Radiological characteristics were correlated with intraoperative hemodynamic instability and surgical approach.

Functional Imaging

Functional imaging is indispensable in hereditary, multifocal, recurrent, or metastatic PPGL [1,3]. Imaging selection is increasingly guided by genotype-specific tumor biology.

Historically, ^123I-metaiodobenzylguanidine (MIBG) scintigraphy was the standard functional modality; however, its sensitivity is limited, particularly in SDHB-related and metastatic disease [12].

Positron emission tomography/computed tomography (PET/CT) using modern radiotracers has significantly improved diagnostic performance:

- ^18F-FDG PET/CT demonstrates high sensitivity in metastatic and SDHB-associated tumors, reflecting increased glycolytic activity [13].
- ^68Ga-DOTATATE PET/CT has emerged as a highly sensitive modality for lesion detection, particularly in SDHx-related tumors, due to strong somatostatin receptor expression [14].
- ^18F-FDOPA PET/CT is particularly useful in selected hereditary and head and neck paragangliomas [3].
- Current evidence supports individualized imaging selection based on:

- germline mutation status,
- biochemical secretion profile,
- tumor location,
- suspicion of metastatic disease [1,3,14].

Functional imaging findings may significantly alter surgical strategy by identifying multifocal disease, occult metastases, or extra-adrenal lesions, thereby reducing the risk of incomplete resection and inappropriate minimally invasive surgical selection.

Preoperative Optimization: Mitigating Catecholamine Risk

Surgical resection of PPGL remains uniquely associated with significant intraoperative hemodynamic instability. Hypertensive crises during tumor manipulation, profound hypotension following adrenal vein ligation, arrhythmias, and stress-induced cardiomyopathy represent persistent perioperative risks [1,3,15]. Despite advances in anesthesia and minimally invasive techniques, meticulous preoperative optimization remains the cornerstone of safe surgical management.

Risk Stratification

Several factors have been consistently associated with increased perioperative instability:

- Tumor size (>5–6 cm)
- Elevated preoperative plasma or urinary catecholamine levels
- Prolonged operative duration
- Catecholamine-induced cardiomyopathy
- SDHB-associated tumors with aggressive biological behavior

Larger tumors and higher catecholamine burden correlate with greater intraoperative blood pressure fluctuations and increased vasopressor requirements [3,16]. Pre-existing cardiomyopathy further amplifies perioperative cardiovascular risk [15].

Optimal outcomes are consistently reported in high-volume endocrine surgery centers with experienced multidisciplinary teams, including endocrine surgeons, anesthesiologists familiar with PPGL physiology, endocrinologists, and intensive care specialists [3,5].

Pharmacologic Preparation

Preoperative alpha-adrenergic blockade remains the foundation of perioperative preparation and should be initiated 10–14 days before surgery [1,3].

Both:

- Non-selective, irreversible α-blockade (phenoxybenzamine)
- Selective α1-adrenergic antagonists (e.g., doxazosin)

are considered effective. Current evidence does not conclusively demonstrate superiority of one regimen over the other in preventing intraoperative hemodynamic instability [17,18].

Beta-blockade may be introduced only after adequate alpha blockade has been achieved to control reflex tachycardia and avoid unopposed alpha-adrenergic stimulation [1].

Preoperative volume expansion through liberal salt intake and fluid administration is essential to reverse chronic catecholamine-induced vasoconstriction and contracted plasma volume, thereby reducing the severity of post-resection hypotension [1,3].

Calcium channel blockers and metyrosine may be considered in selected patients with suboptimal blood pressure control or severe catecholamine excess [3].

Meticulous preoperative preparation significantly reduces perioperative morbidity and remains fundamental to safe tumor resection.

Surgical Management

Fundamental Operative Principles

Regardless of surgical access, several operative principles remain essential:

- Minimal tumor manipulation
- Early identification and ligation of the adrenal vein
- Strict preservation of capsular integrity

Early venous control limits systemic catecholamine surges, while avoidance of capsular disruption reduces the theoretical risk of tumor cell dissemination [19].

Minimally Invasive Adrenalectomy

Minimally invasive adrenalectomy represents the standard of care for localized pheochromocytoma without radiologic evidence of invasion [3,5].

Both transabdominal laparoscopic and posterior retroperitoneoscopic approaches demonstrate excellent perioperative outcomes, including reduced blood loss, shorter hospitalization, and faster recovery compared with open surgery [5,20].

Comparative studies suggest that the posterior retroperitoneoscopic approach may offer:

- Shorter operative time
- Reduced intraoperative blood loss
- Shorter length of stay

in carefully selected patients with smaller tumors [20].

However, large tumors (>7–8 cm), severe obesity, or suspicion of local invasion may necessitate alternative access or open conversion.

Robotic adrenalectomy provides enhanced three-dimensional visualization and improved instrument dexterity. Perioperative outcomes are comparable to conventional laparoscopy, although increased cost and longer setup time must be considered [21].

Institutional Experience: Transabdominal Minimally Invasive Approach

At our institution, a transabdominal laparoscopic approach is employed for all adrenalectomies (Figures 3-5).

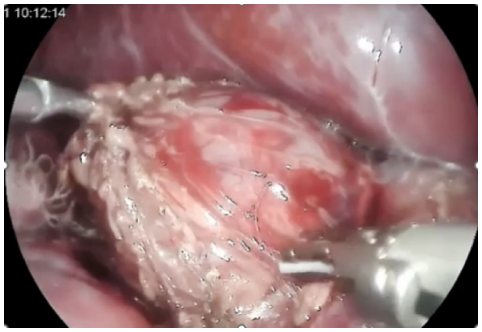

Figure 3: Dissection of the inferior pole of the right adrenal gland.

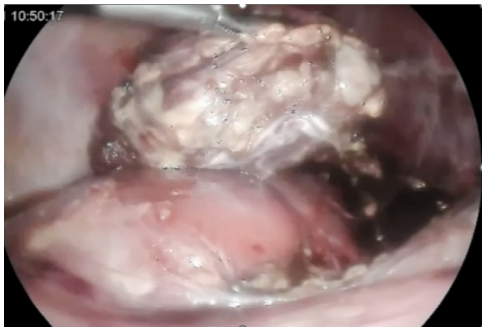

Figure 4: The right adrenal gland mobilized in the paracaval plane with division of the vessels supplying the superior pedicle.

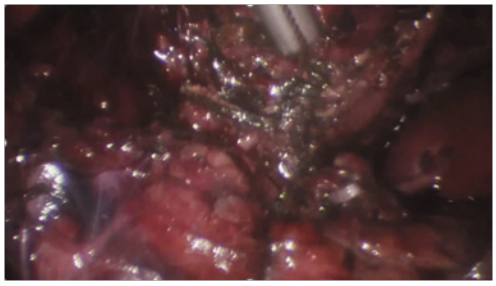

Figure 5: Left adrenal gland. Dissection of the left adrenal pedicle with clip ligation of the vessels supplying the inferior pole.

The patient is positioned in full lateral decubitus (90°), allowing gravity-assisted retraction of adjacent organs.

For left adrenalectomy, three subcostal trocars are placed. For right adrenalectomy, three subcostal trocars and an additional epigastric port are inserted parallel to the inferior liver margin.

Left Adrenalectomy

The spleen is mobilized to allow medial displacement of the splenopancreatic complex. Dissection proceeds in the plane between the pancreatic tail and the left adrenal gland. The adrenal vein is identified early and secured.

Right Adrenalectomy

The right triangular ligament is divided to facilitate medial liver retraction. Dissection is performed in the plane between the inferior vena cava and the right adrenal gland.

In all cases, adrenal vessels are meticulously dissected and clipped with minimal tumor manipulation to limit catecholamine release. The specimen is retrieved intact using an endoscopic retrieval bag to prevent capsular rupture or tumor seeding.

Open Adrenalectomy and Oncologic Considerations

Open adrenalectomy remains indicated in:

- Locally invasive tumors
- Large lesions at risk of rupture
- Suspicion of malignancy or nodal involvement
- Selected SDHB-mutated tumors with aggressive imaging features

Open access may facilitate en bloc resection and improved oncologic clearance in selected patients [3,22].

Cytoreductive surgery may be considered in metastatic PPGL for symptom control and reduction of catecholamine burden, although a clear survival benefit has not been definitively established [3].

Cortical-Sparing Adrenalectomy

Cortical-sparing adrenalectomy is particularly relevant in hereditary syndromes such as multiple endocrine neoplasia type 2 and von Hippel–Lindau disease, where bilateral pheochromocytoma is common [3,23].

Preservation of approximately 15–30% of viable adrenal cortex may maintain endogenous steroid production. However, postoperative adrenal insufficiency still occurs in a proportion of patients, and recurrence risk varies across reported series [23,24].

Therefore, the decision to pursue adrenal-sparing surgery requires individualized balancing of:

- Oncologic safety
- Genetic risk profile
- Likelihood of bilateral disease
- Risk of lifelong steroid dependence

Long-term surveillance is mandatory following cortical-sparing procedures.

Institutional Experience

We retrospectively reviewed seven consecutive patients who underwent surgical resection for PPGL at our tertiary referral center.

The cohort included four men and three women, with a median age of 49 years (range: 44–53 years). Preoperative biochemical evaluation demonstrated elevated plasma 3-methoxytyramine levels ranging from 0.3 to 0.6 nmol/L. Germline genetic testing identified pathogenic variants in three patients: two carriers of SDHA mutations and one carrier of an SDHC mutation.

All patients underwent unilateral adrenalectomy using a minimally invasive transabdominal approach. The median tumor size was 5.2 cm (range: 4.5–6.0 cm). Median length of hospital stay was 8 days (range: 7–10 days).

One patient experienced an intraoperative hypertensive crisis, which was promptly controlled with intravenous antihypertensive therapy without further sequelae. No postoperative complications were recorded, and there was no perioperative mortality.

Among patients who underwent cortical-sparing adrenalectomy, no early postoperative adrenal insufficiency requiring long-term steroid replacement was observed during the available follow-up period.

At a median follow-up of 11 months (range: 10–12 months), no local recurrence or metastatic progression was documented.

Although limited by small sample size and retrospective design, our experience reinforces key principles of contemporary PPGL management: rigorous preoperative optimization, genotype-informed operative planning, disciplined adherence to minimally invasive surgical principles, and structured postoperative surveillance—particularly in patients with hereditary predisposition.

These outcomes align with previously published series demonstrating the safety and feasibility of minimally invasive adrenalectomy in appropriately selected patients [1-3], as well as acceptable endocrine and oncologic outcomes following cortical-sparing procedures [4-6].

Postoperative Management and Surveillance

Immediate postoperative care focuses on hemodynamic stabilization and early detection of complications [1].

Hypotension is common following tumor resection due to abrupt catecholamine withdrawal, persistent alpha-adrenergic blockade, and relative hypovolemia. Management includes:

- Careful intravenous fluid resuscitation
- Temporary vasopressor support when indicated

Close blood pressure monitoring in a high-dependency or intensive care setting is recommended during the early postoperative period, particularly in patients with large tumors or significant preoperative catecholamine excess [1,16].

Postoperative hypoglycemia, attributed to rebound hyperinsulinemia following removal of catecholamine excess, necessitates routine glucose monitoring during the first 24 hours after surgery [1].

Long-Term Surveillance

Long-term follow-up is mandatory for all patients, as all PPGLs are currently classified as tumors with metastatic potential according to the WHO classification [2].

Surveillance strategies typically include:

- Annual biochemical testing (plasma free metanephrines or urinary fractionated metanephrines)
- Periodic cross-sectional or functional imaging in high-risk patients
- Lifelong follow-up in hereditary cases or carriers of high-risk mutations (e.g., SDHB)

European and North American guidelines recommend at least 10 years of follow-up for all patients, with lifelong monitoring in genetically predisposed individuals and those with high-risk tumor biology [1,7].

Structured surveillance is essential not only for early detection of recurrence or metastatic disease but also for long-term cardiovascular risk assessment in patients previously exposed to chronic catecholamine excess.

Morbidity of Untreated Disease

Untreated pheochromocytoma is associated with substantial cardiovascular morbidity and mortality.

Historical autopsy series reported that up to 71% of deaths in previously undiagnosed cases were attributable to cardiovascular complications [26]. Contemporary epidemiological studies continue to demonstrate significantly increased rates of:

- Myocardial infarction
- Stroke
- Arrhythmias
- Heart failure

prior to tumor diagnosis [27].

Chronic catecholamine excess exerts direct and indirect cardiotoxic effects, leading to:

- Reversible or irreversible cardiomyopathy
- Myocardial fibrosis
- Endothelial dysfunction
- Electrical instability

Catecholamine-induced cardiomyopathy may clinically resemble Takotsubo syndrome and can present with acute heart failure, cardiogenic shock, or malignant arrhythmias [15].

Prolonged exposure to supraphysiologic catecholamine levels results in cumulative myocardial injury and may confer persistent cardiovascular risk even after successful tumor resection in a subset of patients [15,27].

These data underscore the imperative of early biochemical diagnosis, timely referral to specialized centers, and definitive surgical management to prevent potentially irreversible cardiovascular damage.

Outcomes Following Cortical-Sparing Adrenalectomy

Cortical-sparing adrenalectomy aims to preserve endogenous adrenal function while maintaining oncologic safety, particularly in hereditary syndromes such as MEN2 and VHL [23,24].

Reported recurrence rates vary considerably across published series. In MEN2-associated pheochromocytoma, gland-based

recurrence rates of up to 51.8% at 10 years have been described in some cohorts. In contrast, other studies report substantially lower recurrence rates ranging from 3% to 10%, depending on duration of follow-up, extent of cortical preservation, and surgical technique [23-25].

Despite preservation of adrenal cortex, postoperative steroid dependence remains clinically significant. Rates of long-term glucocorticoid replacement therapy of up to 43% have been reported, reflecting variability in residual functional cortical tissue and differences in operative strategy [24,25].

Collectively, these data highlight that cortical-sparing adrenalectomy represents a carefully calibrated balance between:

- Functional preservation
- Risk of local recurrence
- Underlying genetic background
- Patient age and probability of bilateral disease

Accordingly, meticulous patient selection, comprehensive preoperative counseling, and lifelong biochemical surveillance are essential following adrenal-sparing procedures.

Discussion

The surgical management of pheochromocytoma and paraganglioma (PPGL) has evolved toward a precision-based paradigm integrating molecular genetics, advanced functional imaging, structured perioperative optimization, and minimally invasive operative techniques. Our findings align with contemporary evidence supporting minimally invasive adrenalectomy as the preferred approach for localized adrenal pheochromocytoma, while reaffirming the continued role of open surgery in selected high-risk scenarios, including large tumors, suspected local invasion, and complex extra-adrenal paragangliomas.

Genetic stratification has emerged as a central determinant of operative planning. Germline mutations—particularly those associated with hereditary syndromes—directly influence recurrence risk, probability of bilateral disease, and long-term metastatic potential. Surgical decision-making therefore extends beyond anatomical resectability to incorporate molecular risk profiling. In carefully selected hereditary cases, cortical-sparing adrenalectomy may reduce the likelihood of lifelong steroid dependence; however, this strategy requires rigorous patient selection and long-term surveillance given the variable recurrence rates and incomplete functional adrenal recovery reported in the literature.

Perioperative hemodynamic instability remains a defining challenge in PPGL surgery. Standardized preoperative alpha-adrenergic blockade, volume expansion, and coordinated multidisciplinary anesthetic management are essential for minimizing cardiovascular complications. Our institutional experience, although limited in size, reinforces the importance of meticulous preparation and treatment within specialized centers to optimize perioperative safety.

Despite favorable short-term outcomes associated with minimally invasive approaches, long-term oncologic data remain heterogeneous across published series, particularly in genetically predisposed populations. Prospective multicenter studies and genotype-specific outcome analyses are needed to further refine risk-adapted surgical algorithms and clarify the role of function-preserving strategies in hereditary disease.

Limitations

This study is limited by its retrospective design and the relatively small cohort size, reflecting the rarity of PPGL. The limited sample restricts statistical power for robust evaluation of long-term oncologic endpoints, including recurrence and disease-specific survival.

In addition, comprehensive genetic testing was not uniformly available for all patients, reflecting the temporal evolution of diagnostic protocols during the study period.

Despite these limitations, the present analysis provides clinically relevant real-world data illustrating contemporary surgical strategy, perioperative optimization, and genotype-informed decision-making within a tertiary referral center.

Conclusions

Surgical resection remains the only definitive treatment for pheochromocytoma and paraganglioma. Contemporary management has shifted toward a precision-oriented paradigm integrating genetic stratification, individualized imaging algorithms, structured preoperative optimization, and tailored operative technique.

Multidisciplinary expertise and treatment within experienced centers are critical determinants of perioperative safety and oncologic control.

Ongoing prospective and genotype-specific outcome studies are required to further refine risk-adapted surgical strategies and optimize functional preservation in hereditary disease.

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