Pheochromocytoma Rule of 10s: A Multidisciplinary Adrenal Center Experience

Richard Aldan, MD; Jason Zheng; Mohamed Nofal, MD; Jaine McKenzie, MD; L. Renee Hilton, MD; Aaron Bolduc, MD

Department of Surgery, Medical College of Georgia at Augusta University

INTRODUCTION

- The "Rule of 10s" (ROT) is historically used to characterize Pheochromocytomas (PCC) as 10% malignant, 10% bilateral, 10% extra-adrenal, and 10% familial.
- Advancements in diagnostic capabilities necessitate a modern re-evaluation of these historic benchmarks.
- This study aims evaluate ROT using our multidisciplinary adrenal center database.

METHODS

- Retrospective review of 31 patients (2018-2025) undergoing adrenalectomy for PCC
- Patients stratified by familial status and malignant status.
- Tumor characteristics, perioperative variables, and postoperative outcomes were compared between cohorts.

RESULTS

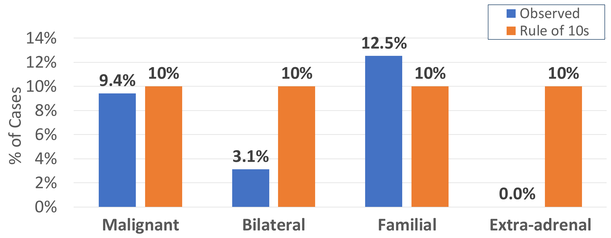

Figure 1. Observed pheochromocytoma characteristics compared with the Rule of 10s (n=31).

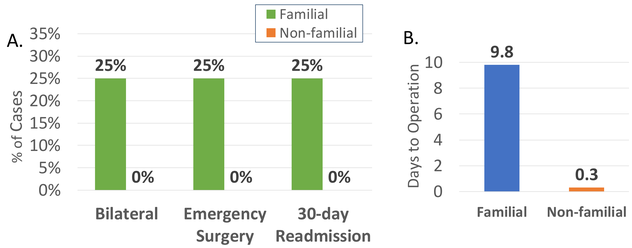

Figure 2. Clinical outcomes (A) and mean days to operation (B) by familial status (Non-familial n=2; Familial n=4).

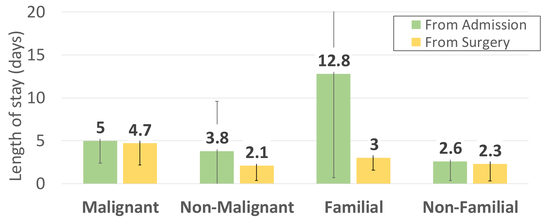

Figure 3. Mean ± SD length of stay from admission and from time of surgery by malignancy and familial status in PCC patients.

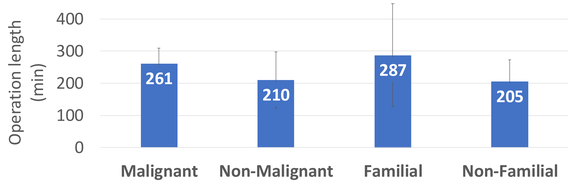

Figure 4. Mean ± SD length of operation in PCC patients by malignancy and familial status.

- Familial PCCs were associated with longer time to operation and higher rates of bilateral tumors, emergent operations, and 30-day readmissions (Fig. 2).
- Malignant PCCs were associated with longer length of stay (Fig. 3) and trended towards longer operative times (Fig.4) and open surgery
- Malignant PCC was also associated with increased preoperative dialysis, higher preoperative creatinine, lower preoperative hematocrit, and more frequently required ≥ 4 antihypertensive medications.

CONCLUSIONS

- Familial PCCs may be more common than previously thought, while malignant, bilateral, and extra-adrenal PCCs may be less common.
- Familial and malignant PCCs have a higher propensity for perioperative complexity and suboptimal outcomes.
- As diagnostic and genetic testing continue to evolve, adrenal centers should maintain high index of suspicion and close perioperative monitoring for familial and malignant PCC.

REFERENCES

Eisenhofer, G., Pamporaki, C., & Lenders, J. W. M. (2023). Biochemical assessment of pheochromocytoma and paraganglioma. Endocrine Reviews, 44(5), 862–909. https://doi.org/10.1210/endrev/bnad011

Lenders, J. W. M., Duh, Q.-Y., Eisenhofer, G., Gimenez-Roqueplo, A.-P., Grebe, S. K. G., Murad, M. H., Naruse, M., Pacak, K., & Young, W. F. (2014). Pheochromocytoma and paraganglioma: An Endocrine Society clinical practice guideline. *Journal of Clinical Endocrinology & Metabolism*, 99(6), 1915–1942. https://doi.org/10.1210/jc.2014-1498
Link: https://academic.oup.com/jcem/article/99/6/1915/2537399

Conzo, G., Pasquali, D., Colantuoni, V., Circelli, L., Tartaglia, E., Gambardella, C., Napolitano, S., Mauriello, C., Avenia, N., Santini, L., & Sinisi, A. A. (2014). Current concepts of pheochromocytoma. *International Journal of Surgery*, 12(5), 469–474. https://doi.org/10.1016/j.ijsu.2014.04.001