

## Functional Outcome After Resection of von Hippel-Lindau Disease-Associated Lumbosacral Hemangioblastomas.

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#### Introduction

Lumbosacral nerve root hemangioblastomas in von Hippel-Lindau disease (VHL) can underlie significant neurologic signs and symptoms. Despite their associated morbidity, their surgical management and the functional outcomes of resection are not well defined. To define the results of surgical management of lumbosacral hemangioblastomas, we analyzed functional outcomes after surgery for these lesions.



Magnetic resonance imaging of a cauda equina hemangioblastoma with associated nerve root cyst. Sagittal T1 -weighted post-contrast (left) and T2weighted imaging demonstrate a contrast-enhancing hemangioblastoma at L2 (white arrows) with an associated cystic mass (white arrowhead). Axial T2-weighted imaging (right) rostral to tumor demonstrates a cystic mass (white arrowhead) displacing the cauda equina anteriorly and laterally.

# Methods

VHL patients that underwent surgical resection of lumbosacral hemangioblastomas at the National Institutes of Health and the University of Virginia were included. Clinical and radiologic follow-up was performed at 6- to 12-month intervals after surgery. Functional outcome was defined using the McCormick clinical grading scale.

#### Results

Fourteen patients (7 male, 7 female) underwent 18 operations for 21 lumbosacral hemangioblastomas. Most patients presented with multiple symptoms including pain (12 patients), numbress (9), urinary complaints (6), and weakness (2). Mean combined preoperative tumor volume was 2.6±3.6 cm3 (range 0.3 to 15.0 cm3). Four tumors at 3 operations were not resected due to a motor nerve root origin. Gross total resection was achieved in 13 (87% of operations when resection was attempted). All tumors originated from within the involved nerve fascicle. Median follow-up was 5.9 years. New neurologic symptoms were noted after 11 operations (61%) but most (7; 64%) resolved within 2 weeks of surgery. At 6 months follow-up, 15 patients were stable, 2 patients were improved, and 1 patient was worse. Thirteen patients (72%) noted improvement of their presenting symptoms.



Pre-operative axial (left) and sagittal (center) T1-weighted post-contrast magnetic resonance imaging demonstrating a large (volume: 15 cm3) cauda equina hemangioblastoma (white arrowheads) involving and eroding the sacrum. Due to the size of this lesion, selective embolization was performed preoperatively. Gross total resection was achieved and no recurrence has been observed through 13 years follow-up (most recent imaging, right).

Histopathologic analysis (10x) of a cauda equina hemangioblastoma demonstrates tumor infiltration (black arrows) of involved nerve root (asterisk) by hematoxylin and eosin stain (H&E; left). Neuron-specific enolase immunohistochemistry (NSE; right) identifies hemangioblastoma tissue (positive staining; top) infiltrating nerve root axons (does not stain normal axons; asterisk).



### Conclusions

VHL-associated lumbosacral hemangioblastomas can be associated with motor nerve roots and may be large and challenging to manage surgically. Despite these findings, most patients improve with surgery, which should remain the primary therapeutic modality for patients with symptomatic lesions.

#### References

Lonser RR, Wait SD, Butman JA, et al. Surgical management of lumbosacral hemangioblastomas in von Hippel-Lindau syndrome. J Neurosurg. 2003; 99 (1 Suppl): 64-9.

#### Learning Objectives

By the conclusion of this session, participants should be able to:

 Understand the typical presentation of lumbosacral hemangioblastomas in VHL

2) Describe the risks and benefits of surgical resection

 Understand the role of surgery in the management of VHL-associated lumbosacral hemangioblastomas