Assessment of intra-abdominal manifestations of Von Hippel-Lindau disease using Magnetic Resonance Imaging; a pictorial review.

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Purpose
The aim of this presentation is to demonstrate the broad spectrum of the intra-abdominal clinical manifestations of Von Hippel-Lindau disease and their characteristic features on Magnetic Resonance Imaging.

Methods and materials
Von Hippel-Lindau disease is a protean, multi-systemic disorder with potentially treatable manifestations. Genetic testing, advanced surgical techniques and screening have significantly reduced morbidity and mortality for these patients, with imaging playing a key role in this multi-disciplinary approach.

We review the abdominal Magnetic Resonance studies of individuals with Von Hippel-Lindau disease and discuss the pertinent findings.

Results
There is a wide spectrum of benign and malignant tumours associated with Von Hippel-Lindau disease. These include renal and pancreatic cysts and tumours, neuroendocrine pancreatic tumours and adrenal pheochromocytomas.

Renal cysts and tumours
The renal lesions vary from simple cysts to more complex cysts or solid lesions (RCC). The lesions are bilateral in up to 75% of patients and renal tumours occur at a younger age in Von Hippel-Lindau disease (mean 30-36 years) compared to the general population.

Pancreatic cysts and tumours
These include simple cysts, serous cystic adenoma and rarely adenocarcinoma and pancreatic neuroendocrine tumours.

Phaeochromocytomas
In Von Hippel Lindau disease phaeochromocytomas are usually bilateral, present in a younger age and can be ectopic. Imaging usually includes MRI and 18F-MIBG.

Liver cysts
They are usually simple and multiple.

Conclusion
Knowledge of the disease spectrum and familiarisation with the imaging characteristics is paramount in image interpretation and early disease detection. Screening protocols depend on the age group. For the intra-abdominal manifestations of VHL a yearly ultrasound scan is advised in the early teenage years, which change to MRI in adulthood.

References
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