Hypoglycaemia unawareness – a challenge in the management of a Von Hippel-Lindau patient

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**Background**

**Von Hippel-Lindau disease**
- Rare genetic disorder
- Mutation VHL gene
- Autosomal dominant pattern

**Benign tumours in multiple organ systems with potential for malignant change**
- Hemangioblastomas (retinal and central nervous system)
- Pheochromocytomas
- Multiple cysts in the pancreas and kidneys
- Increased risk for renal cell carcinoma

**Hypoglycaemia**
- Key physiologic defences:
  - Insulin
  - Secretion of glucose counterregulatory hormones (namely glucagon and epinephrine)

**Hypoglycaemic unawareness:**
- Absence of warning signs of impending neuroglycopenia

**Case Report**

- Normal BMI
- Without relevant medical history
- Without usual medication

**Hypertension**
- Palpitations
- Diaphoresis

**24-hour urinary catecholamine levels**

<table>
<thead>
<tr>
<th>Catecholamine</th>
<th>Value</th>
<th>Reference</th>
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<tbody>
<tr>
<td>Normetanephrine</td>
<td>8304 µg/24h</td>
<td>86 – 444</td>
</tr>
<tr>
<td>Norepinephrine</td>
<td>2511 µg/24h</td>
<td>15 – 80</td>
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<tr>
<td>Vanillylmandelic acid</td>
<td>33.5 mg/24h</td>
<td>1.4 – 6.5</td>
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**Abdominal CT scan:** compatible with bilateral pheochromocytoma

**Bilateral Pheochromocytoma**

**Proximal pancreatectomy**
- (histopathological study: pancreatic neuroendocrine tumours)

**Total pancreateodudenectomy**
- (histopathological study: pancreatic neuroendocrine tumours, chronic pancreatitis and pancreatic cysts)

**Diabetes secondary to pancreatectomy**

**Conclusion**

Von Hippel-Lindau disease is a rare genetic disorder predisposing to a variety of malignant and benign neoplasms.

This case report highlights the difficult and challenging management of hypoglycaemia unawareness in a Von Hippel-Lindau patient previously submitted to bilateral adrenalectomy and total pancreatectomy.