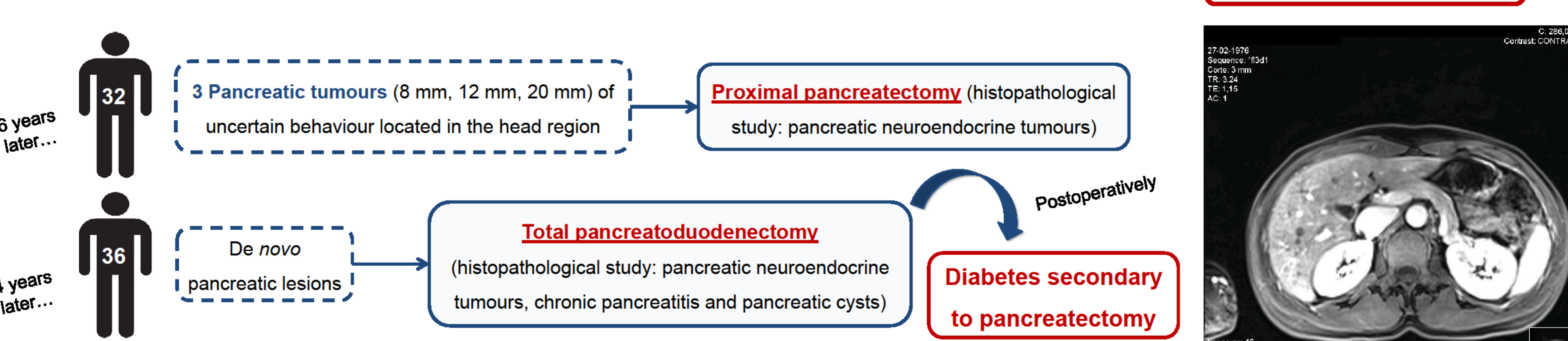
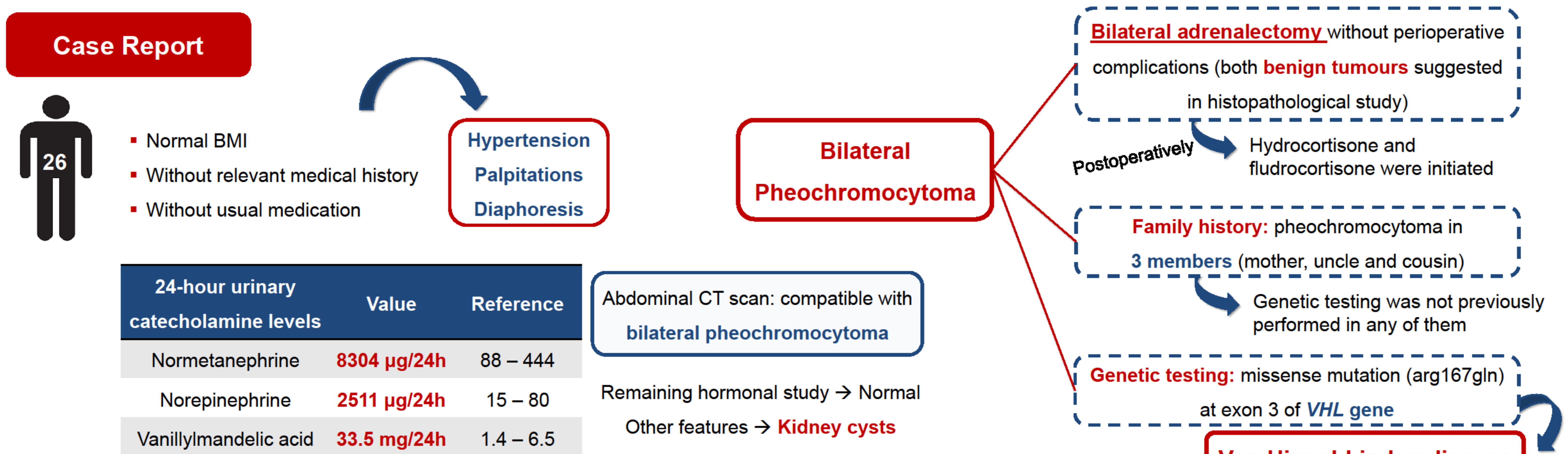
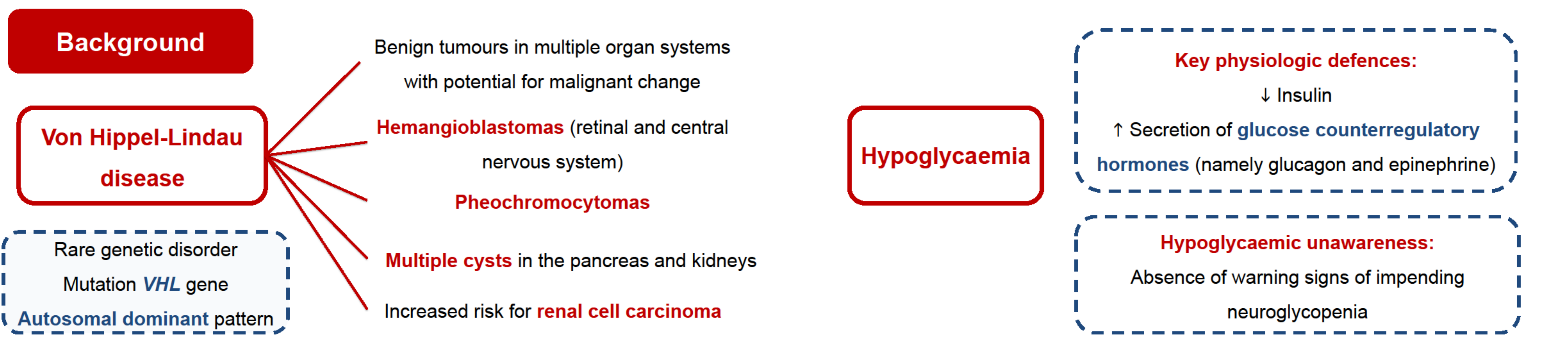


# Hypoglycaemia unawareness – a challenge in the management of a Von Hippel-Lindau patient

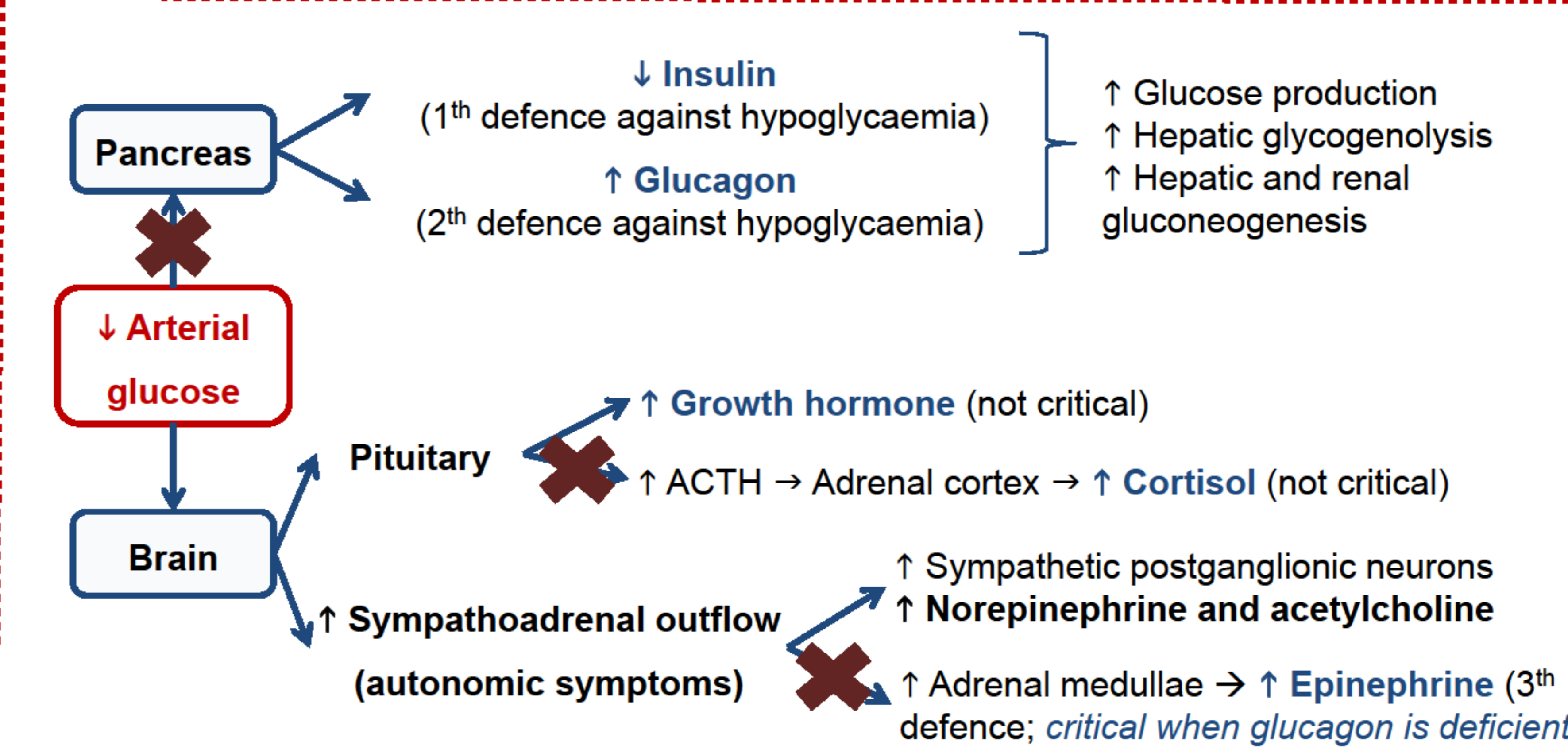
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- He was treated with **multiple daily insulin injection regimen** (detemir twice daily plus prandial lispro), but difficult clinical management.
  - He had **impaired glucose counterregulatory responses**, leading to **severe and recurrent hypoglycaemia unawareness**.
  - These episodes frequently had **severe neuroglycopenic symptoms**, seizures and loss of consciousness requiring glucagon or parenteral glucose therapy.
- Currently... 40
- The patient is learning **carbohydrate counting** with symptoms improvement.
  - He has less stringent glycaemic goals.
  - Use of an **insulin pump** was proposed.



**Conclusion**

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

References: 1. Lonser RR, et al. Von Hippel-Lindau disease. *Lancet* 2003 Jun 14;361(9374):2059-67. 2. Melmed S et al. *Williams Textbook of Endocrinology*, 12th edition. Elsevier (2011). 3. Powers A. Diabetes Mellitus: Complications. In Kasper DL et al, *Harrison's Principles of Internal Medicine*, 19th ed, New York, McGraw-Hill, 2015. 4. Sprague JE, Arbeláez AM. Glucose Counterregulatory Responses to Hypoglycemia. *Pediatr Endocrinol Rev*. 2011 Sep; 9(1): 463-475.