Histopathology of Pancreas in Congenital Hyperinsulinism

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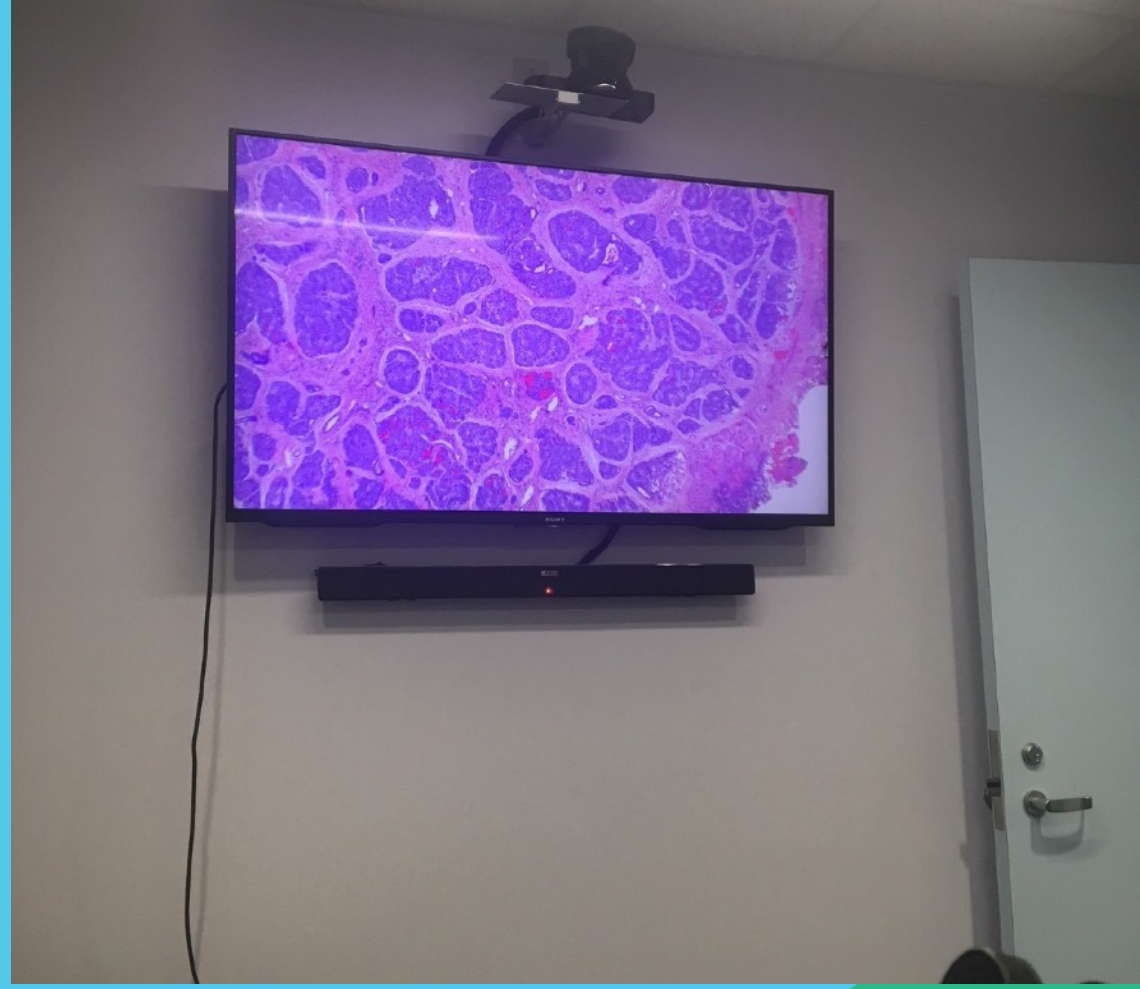
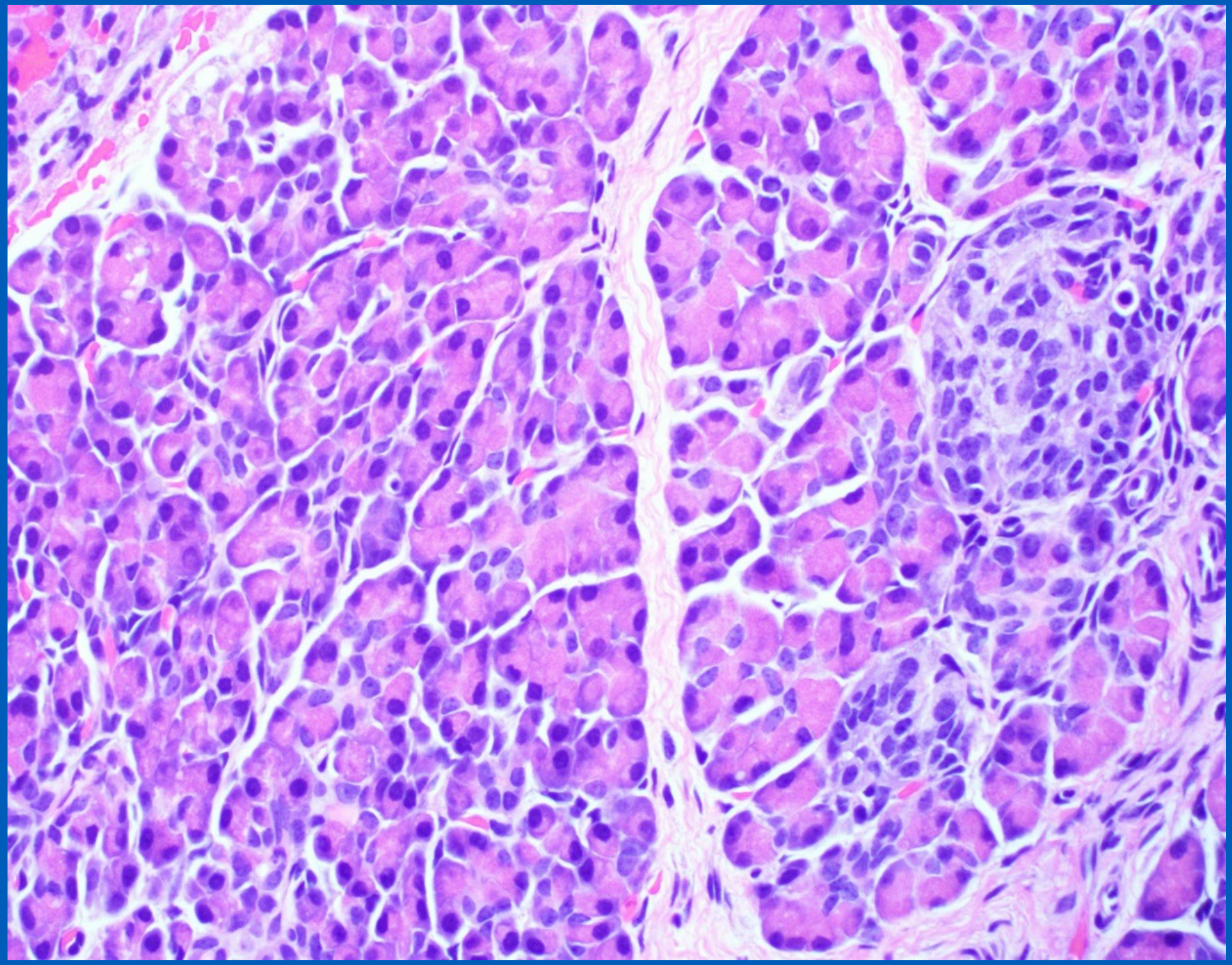
June 1, 2024



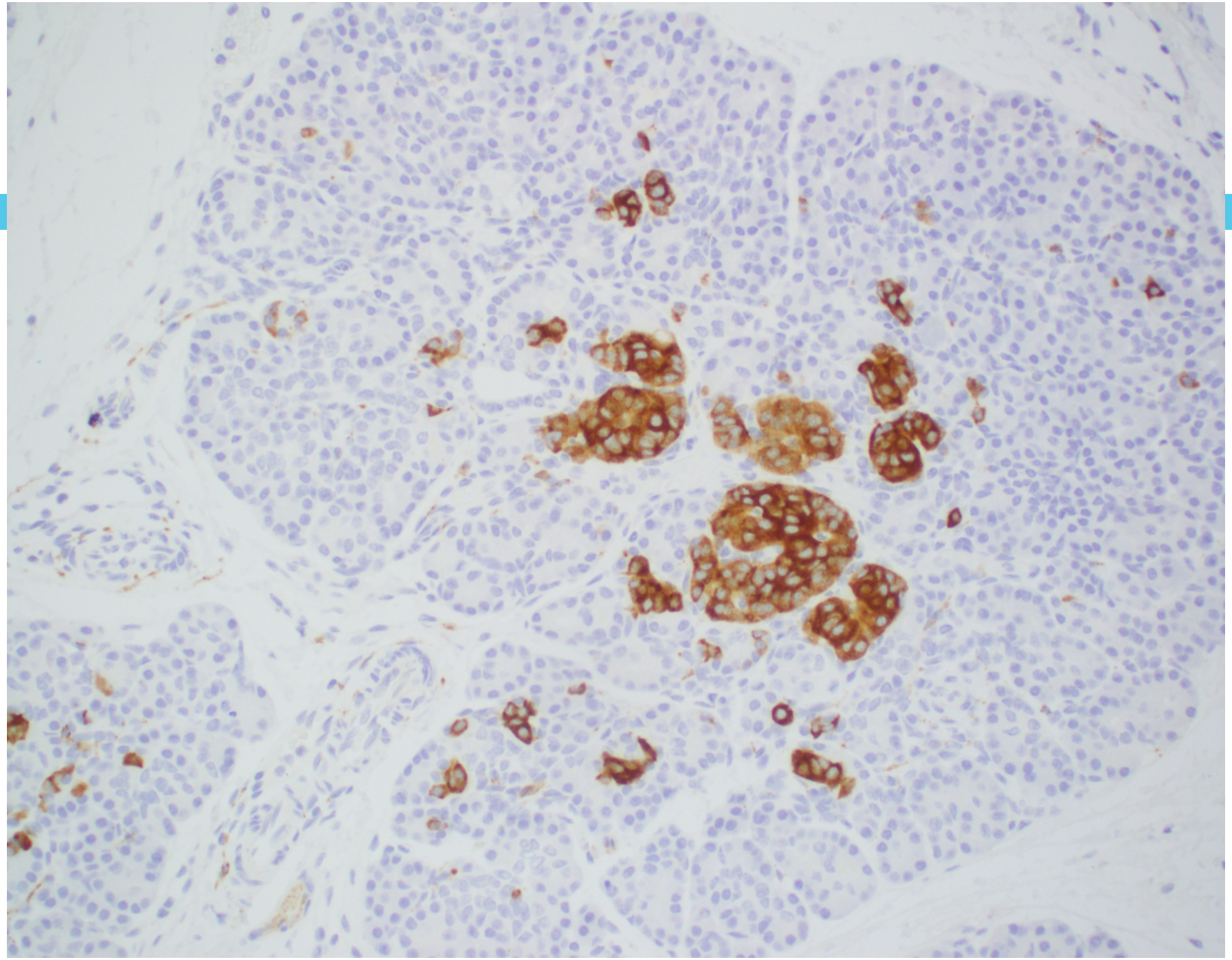
KATP HI – classified into two distinct histologic forms

Genetics	Result of mutation	Response to Diazoxide	Histologic subtype
Biallelic inheritance of *recessive* KATP channel mutations	Absence of plasma membrane KATP channels	Diazoxide unresponsive	Diffuse form
Monoallelic paternally inherited *recessive* KATP channel mutations + loss of maternal chromosome 11p15 (loss of tumor suppressor – lose p57)	Localized absence of plasma membrane KATP channels in pancreas and focal hyperplasia	Diazoxide unresponsive	Focal form
Monoallelic dominant KATP channel mutations	Mutant subunits of channel complex = impaired channel function	Variable responsiveness depending on resultant channel activity	Diffuse form

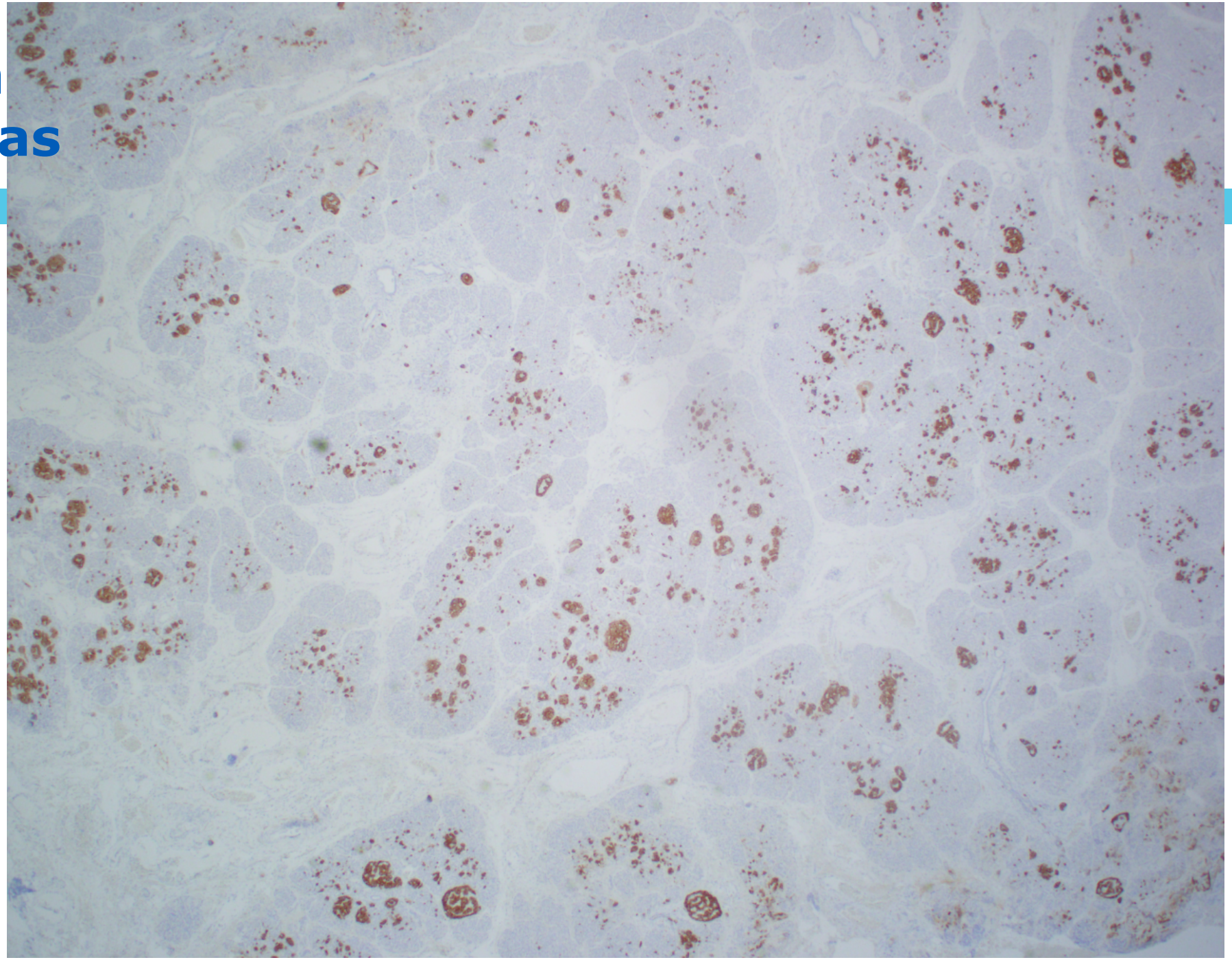
NORMAL PANCREAS



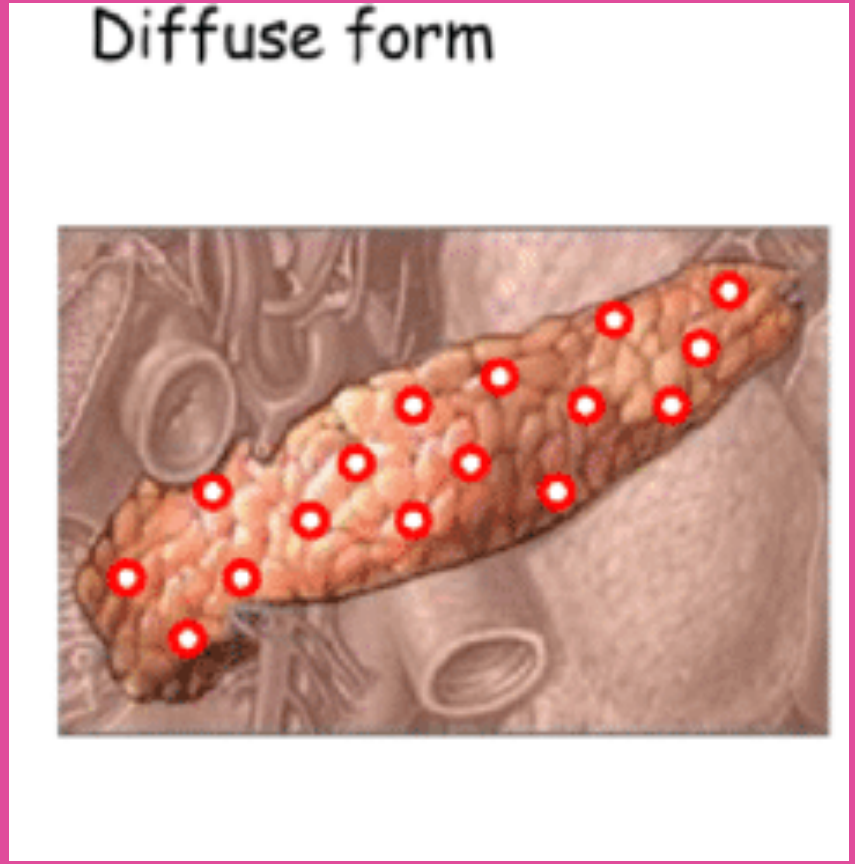
CookChildren's



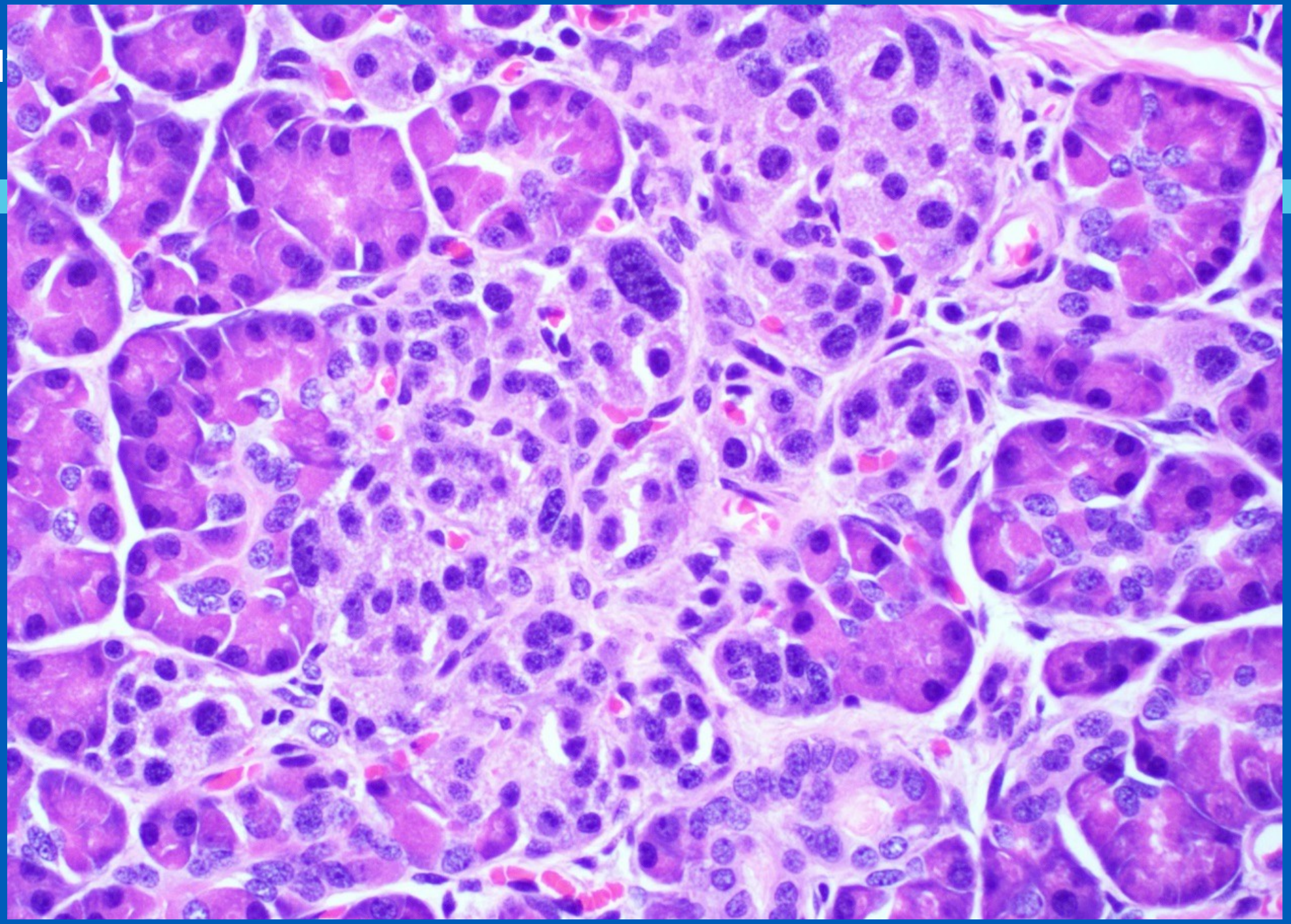
Synaptophysin normal pancreas



DIFFUSE FORM



Nucleomegal



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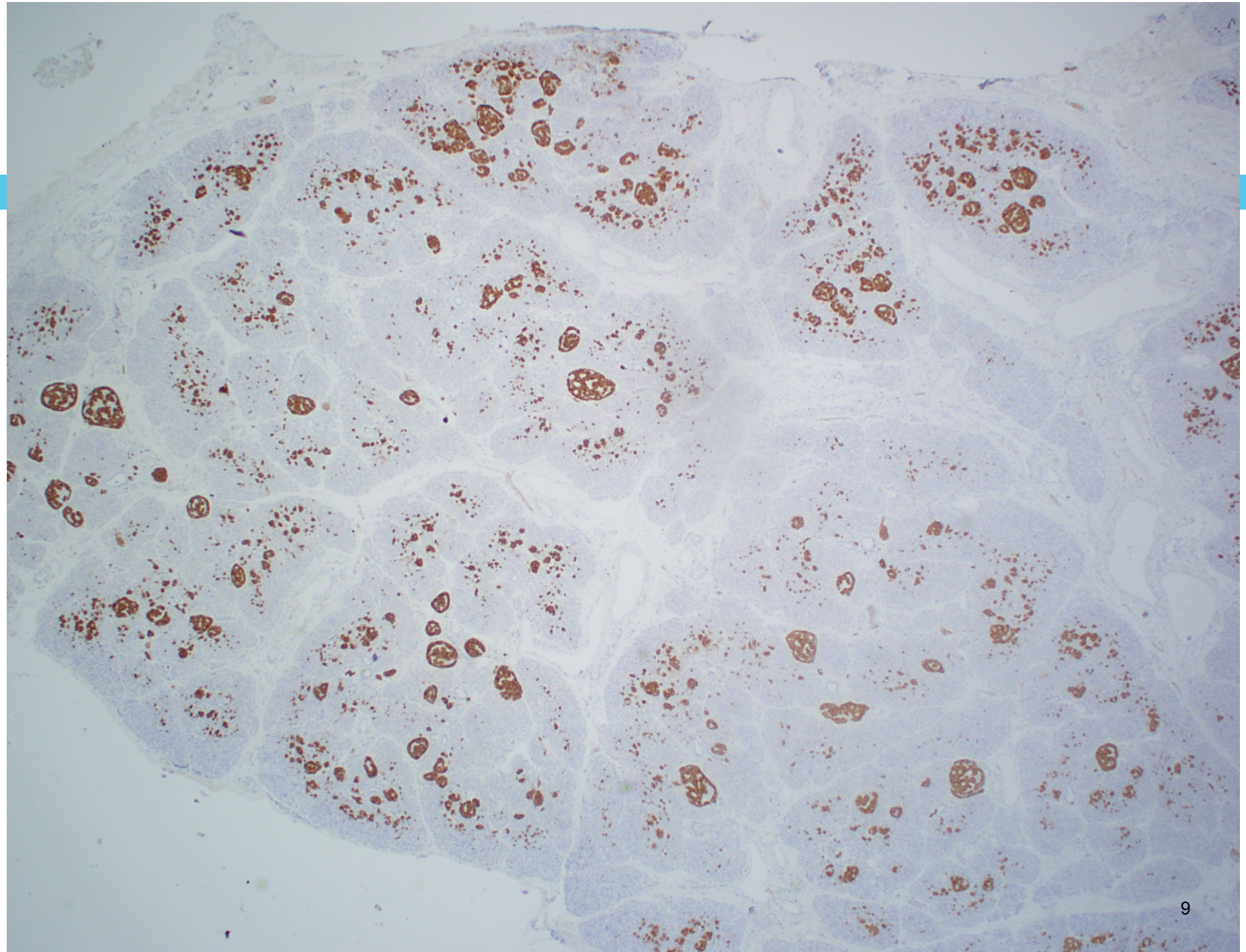
Synaptophysin diffuse form



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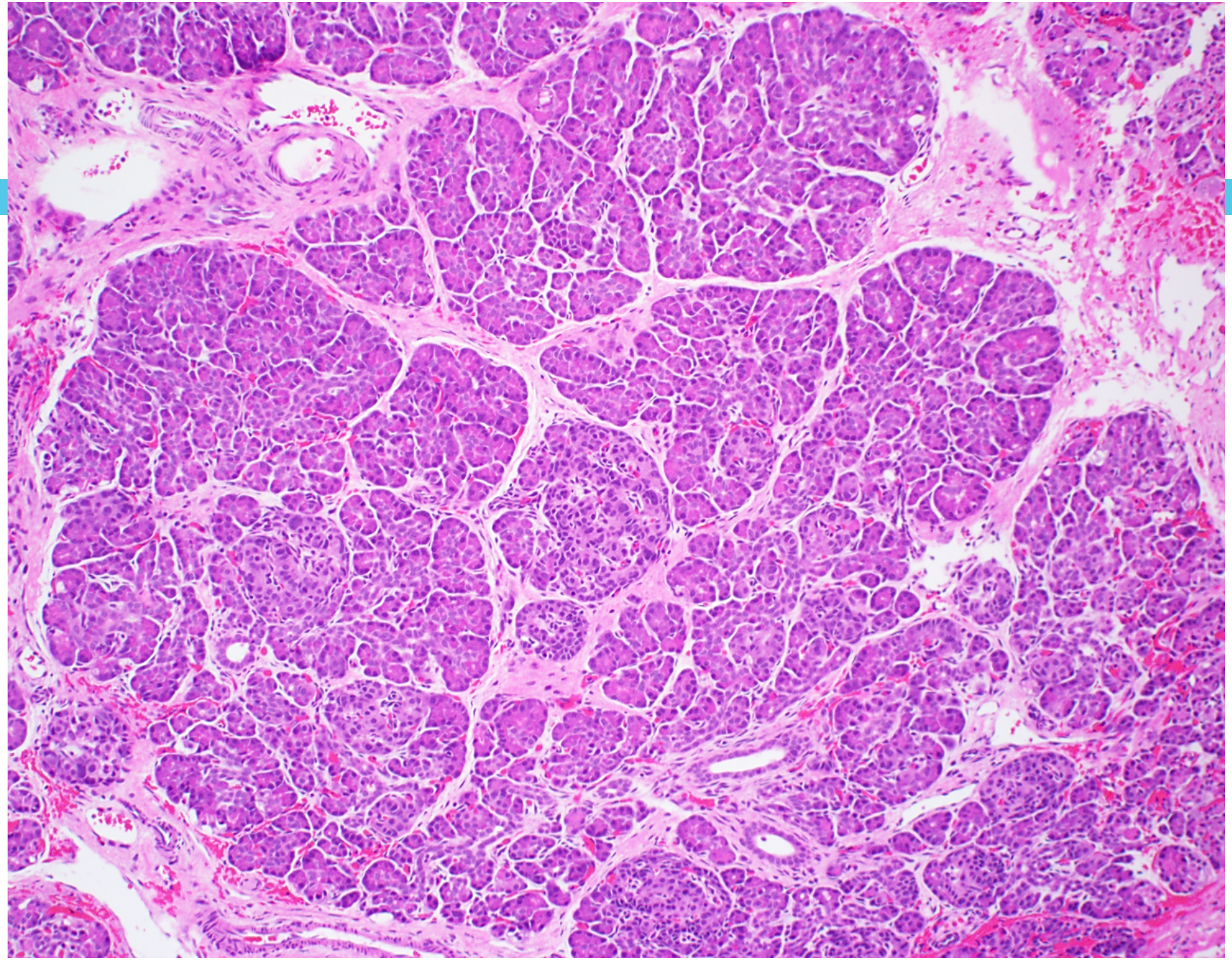

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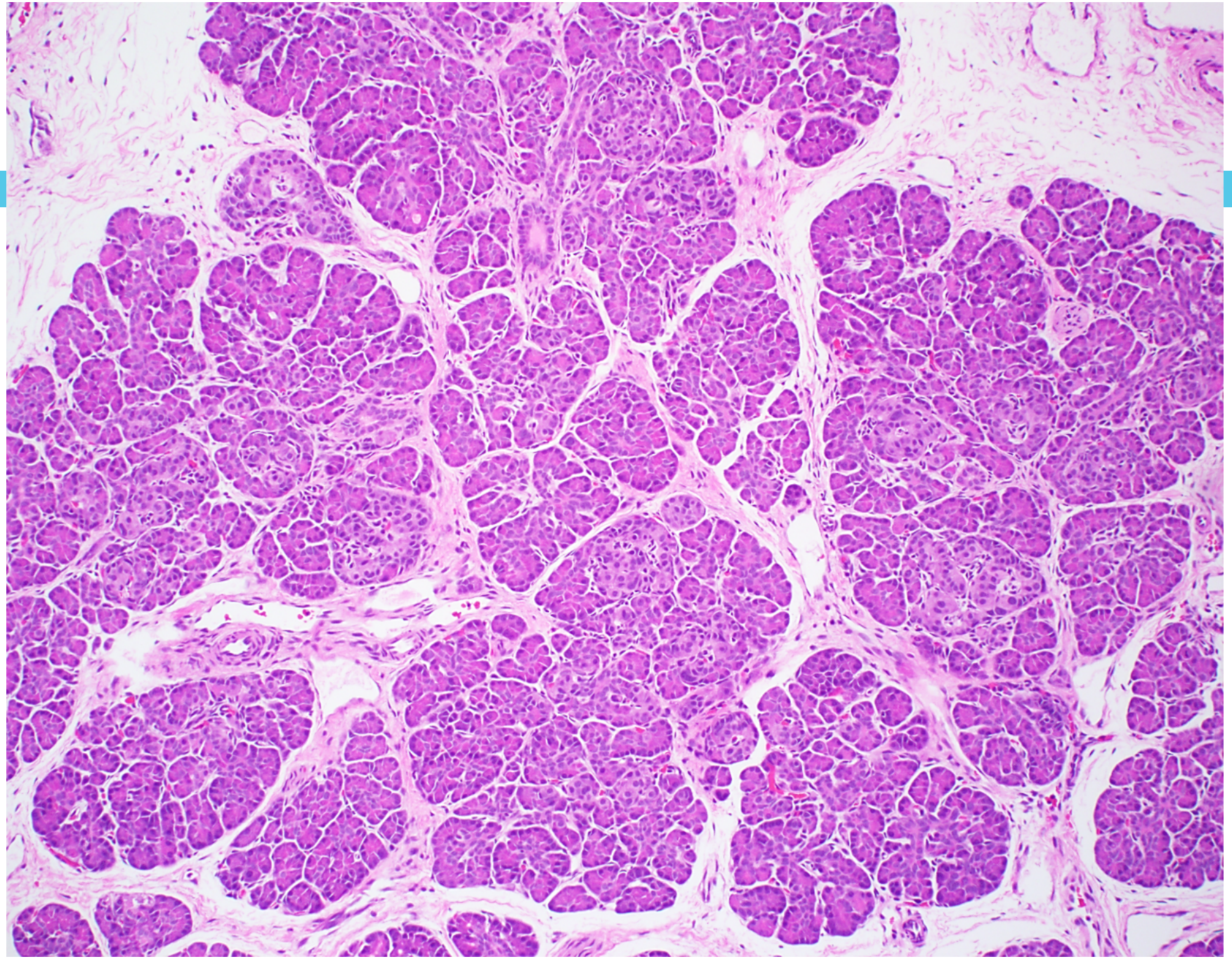

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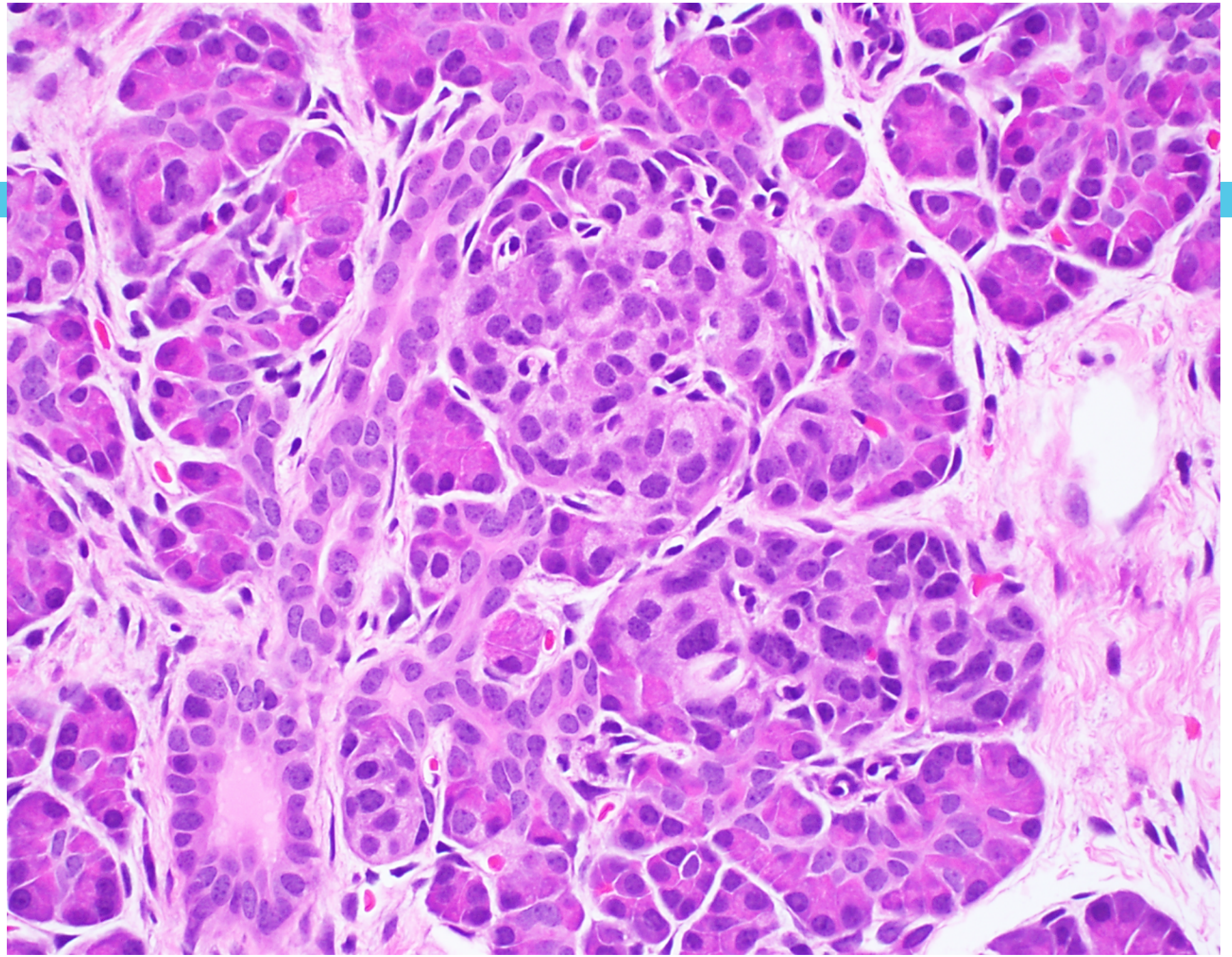

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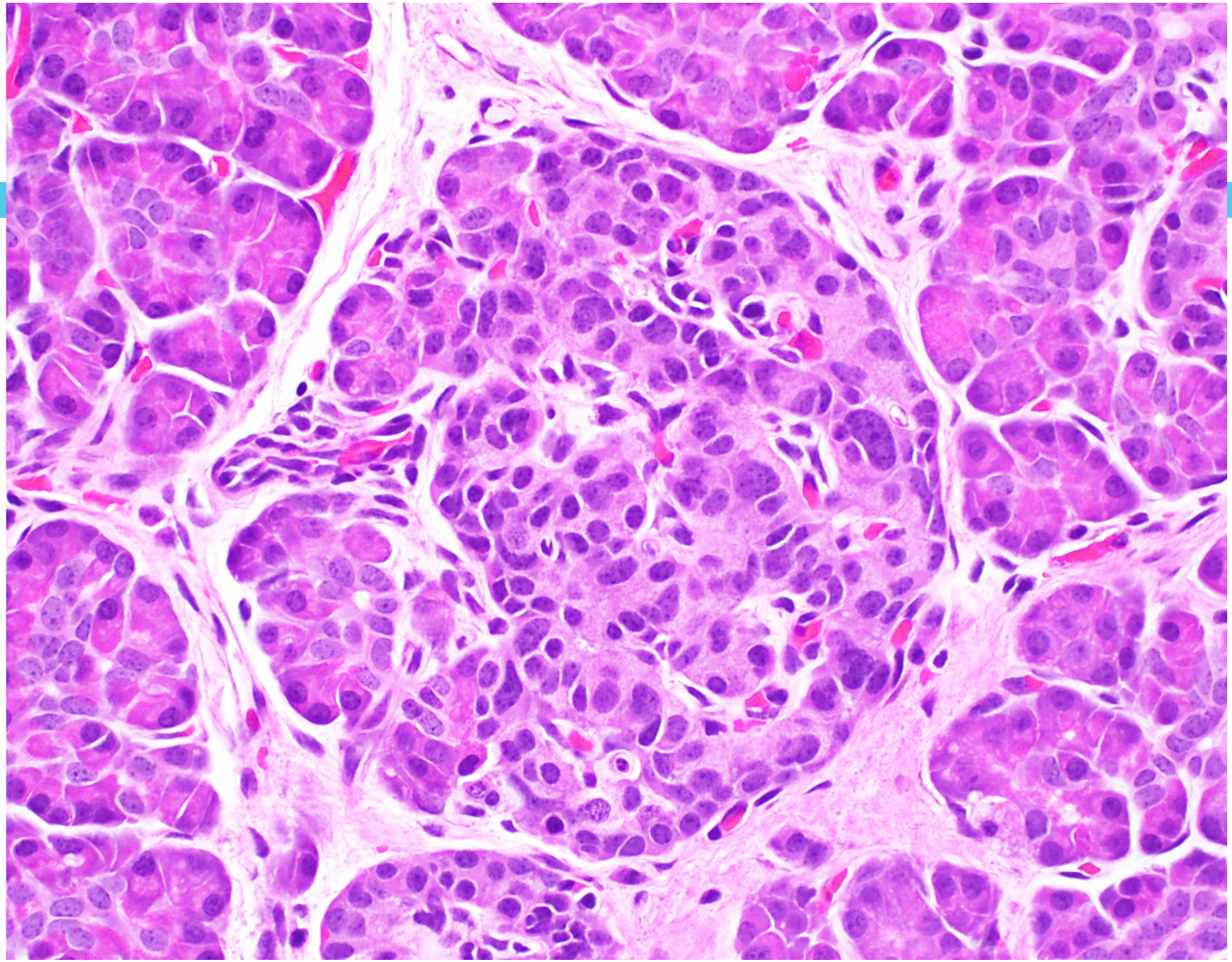

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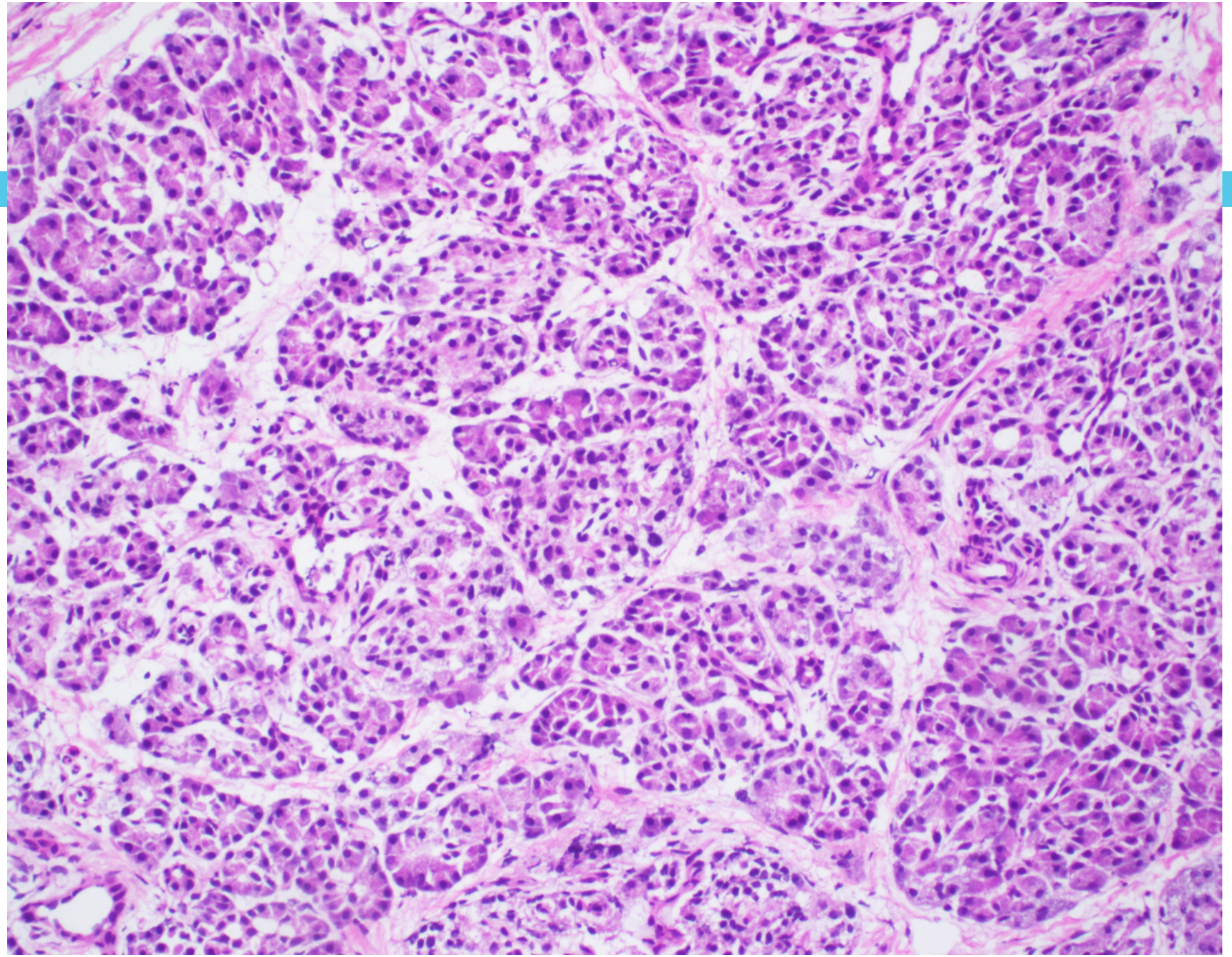
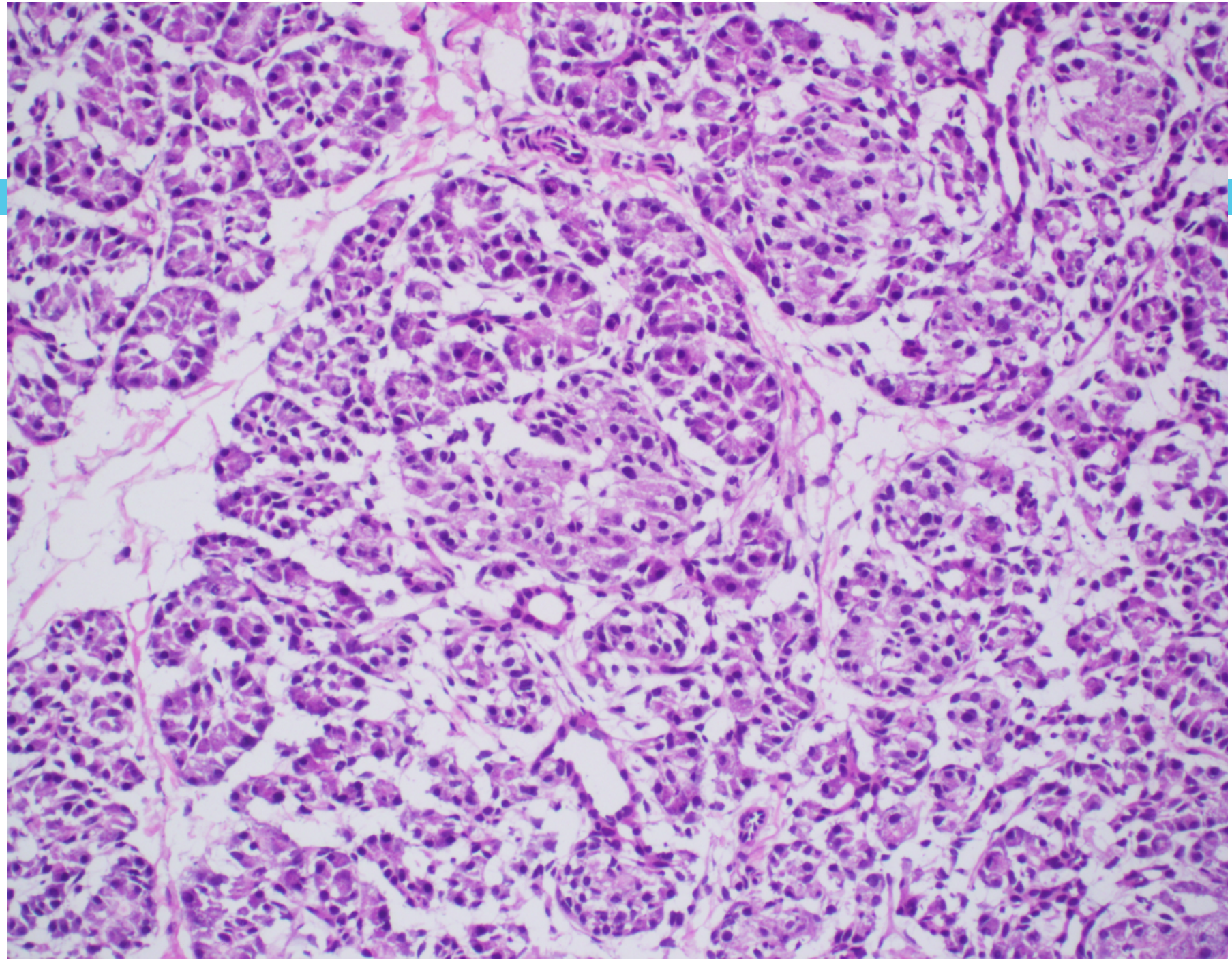
Frozen section histology

Frozen section





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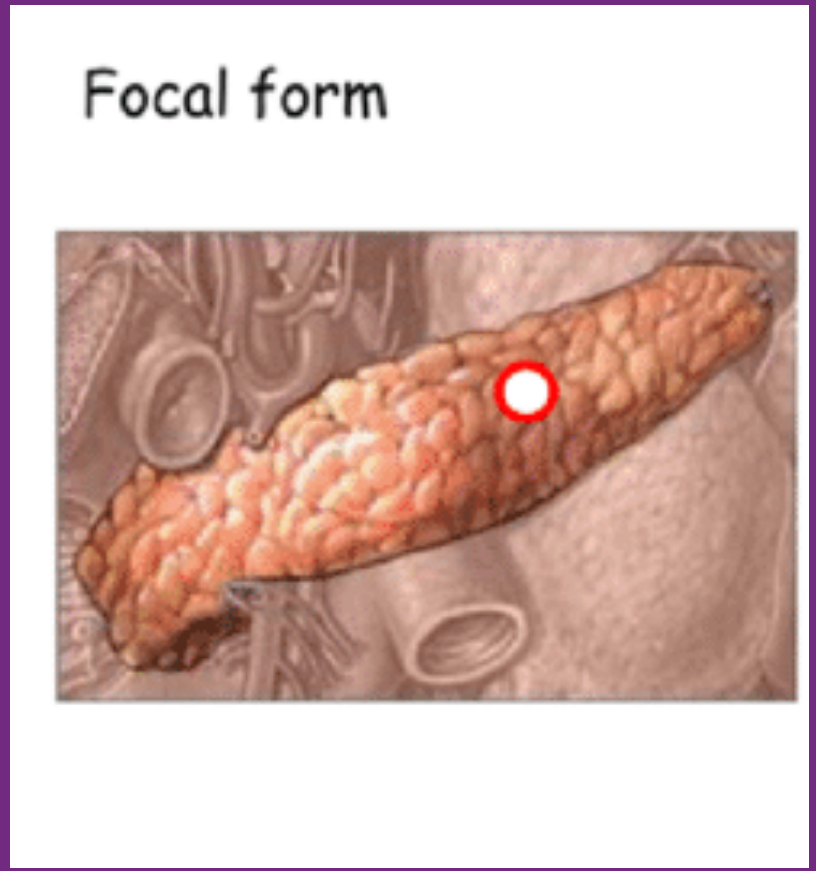
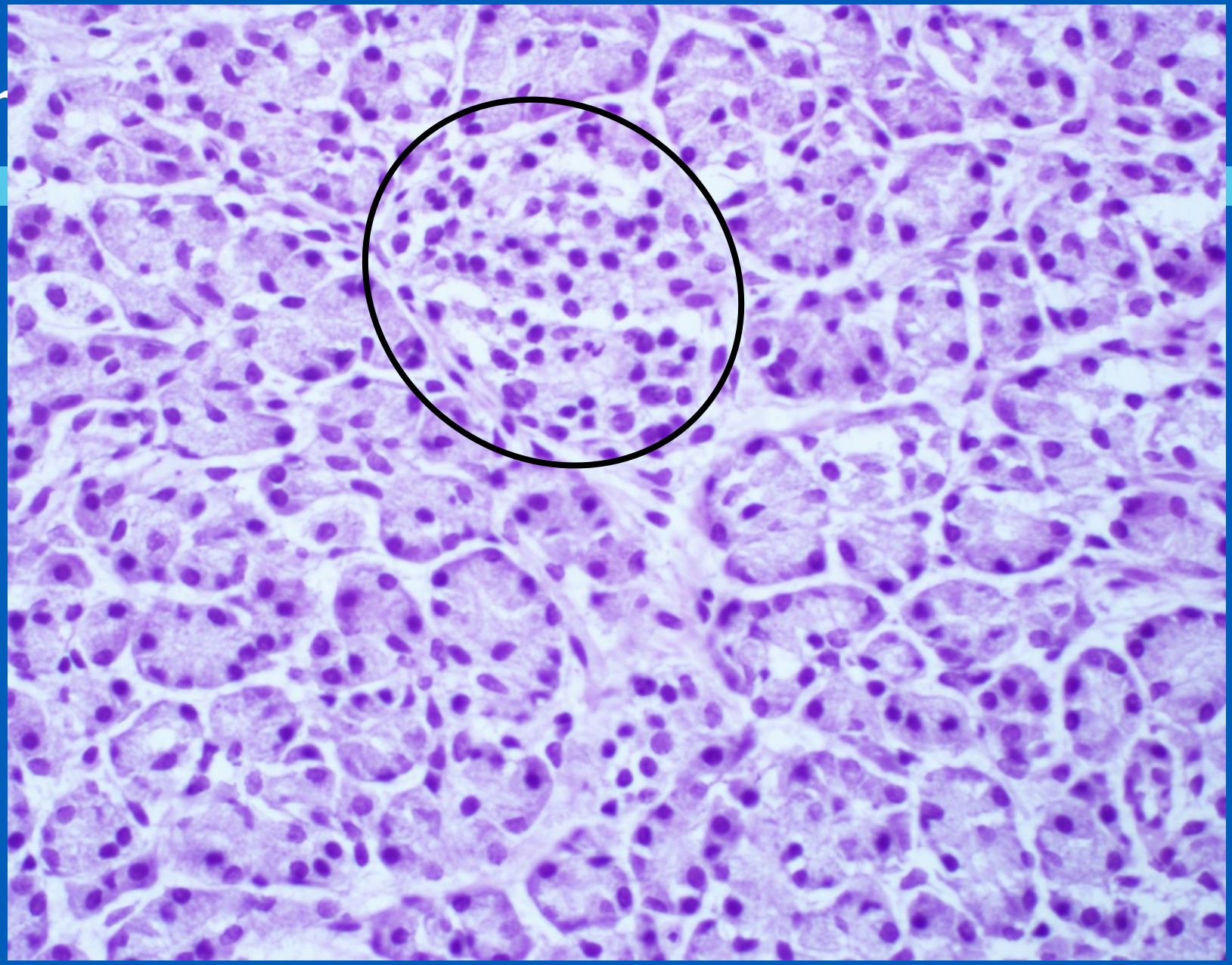
FOCAL FORM



Normal pancreas biopsy frozen sectior



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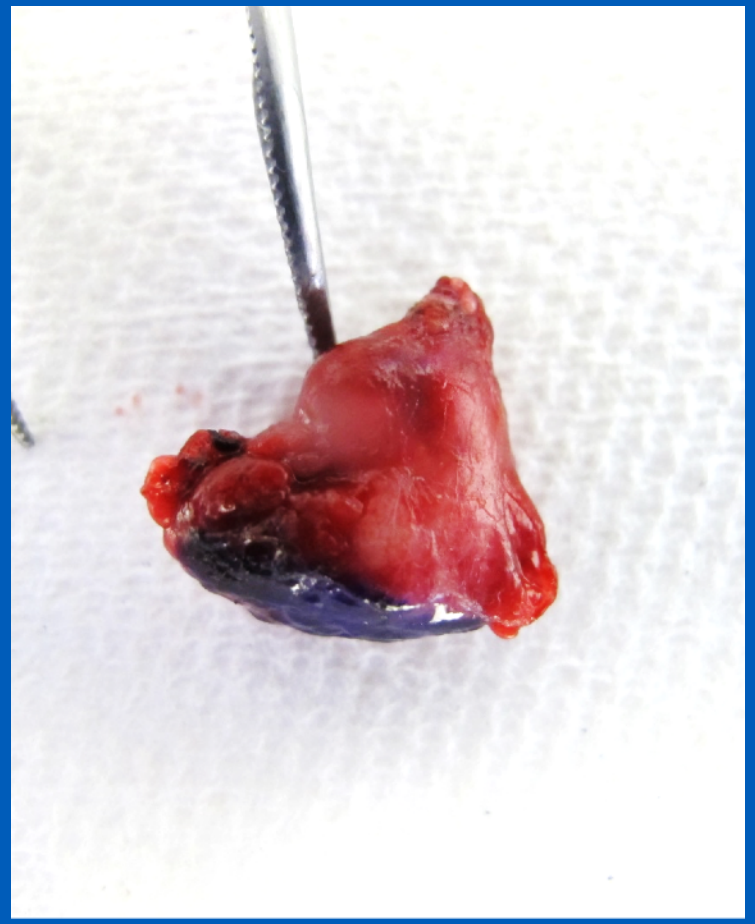
Focal lesion with ink at margin



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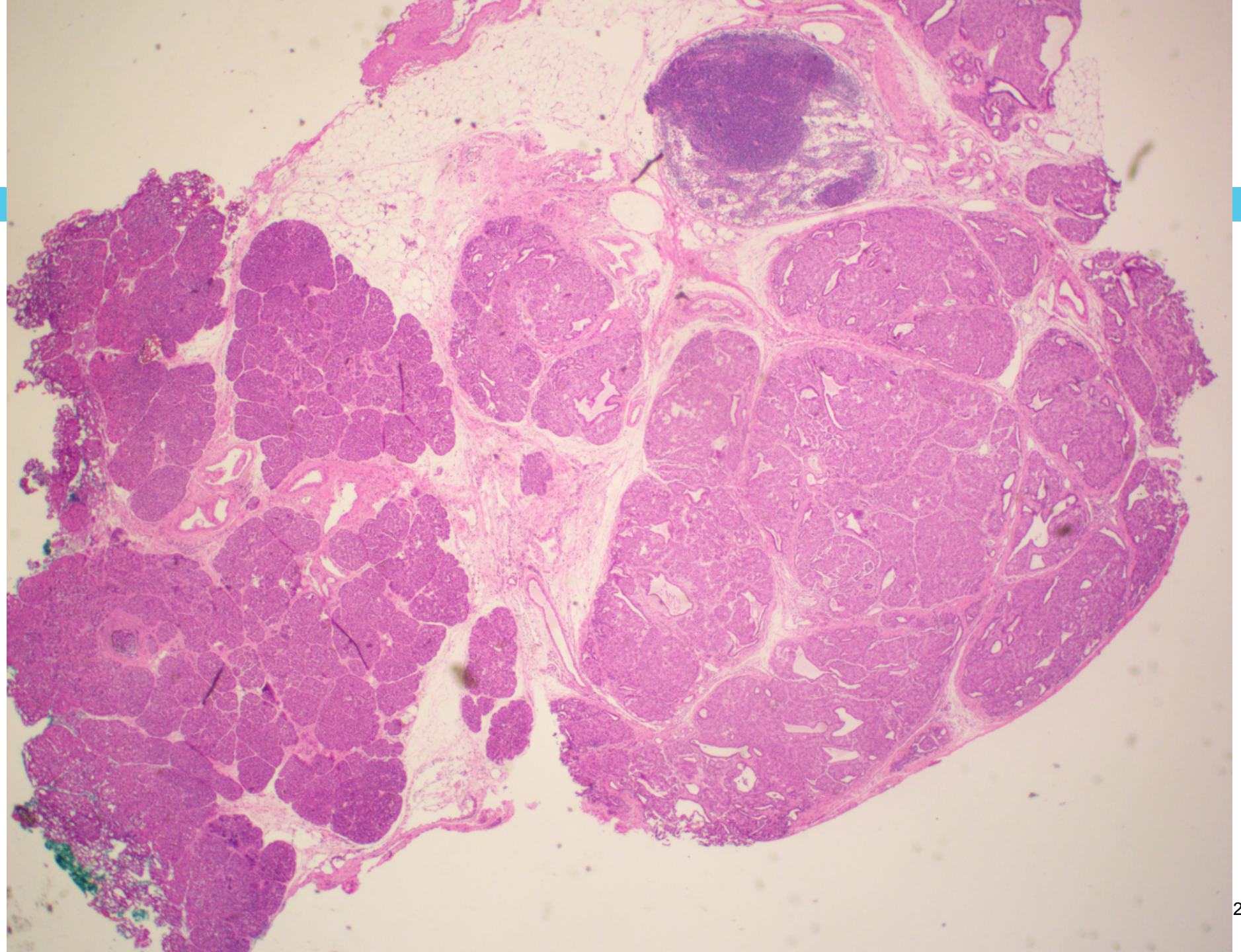

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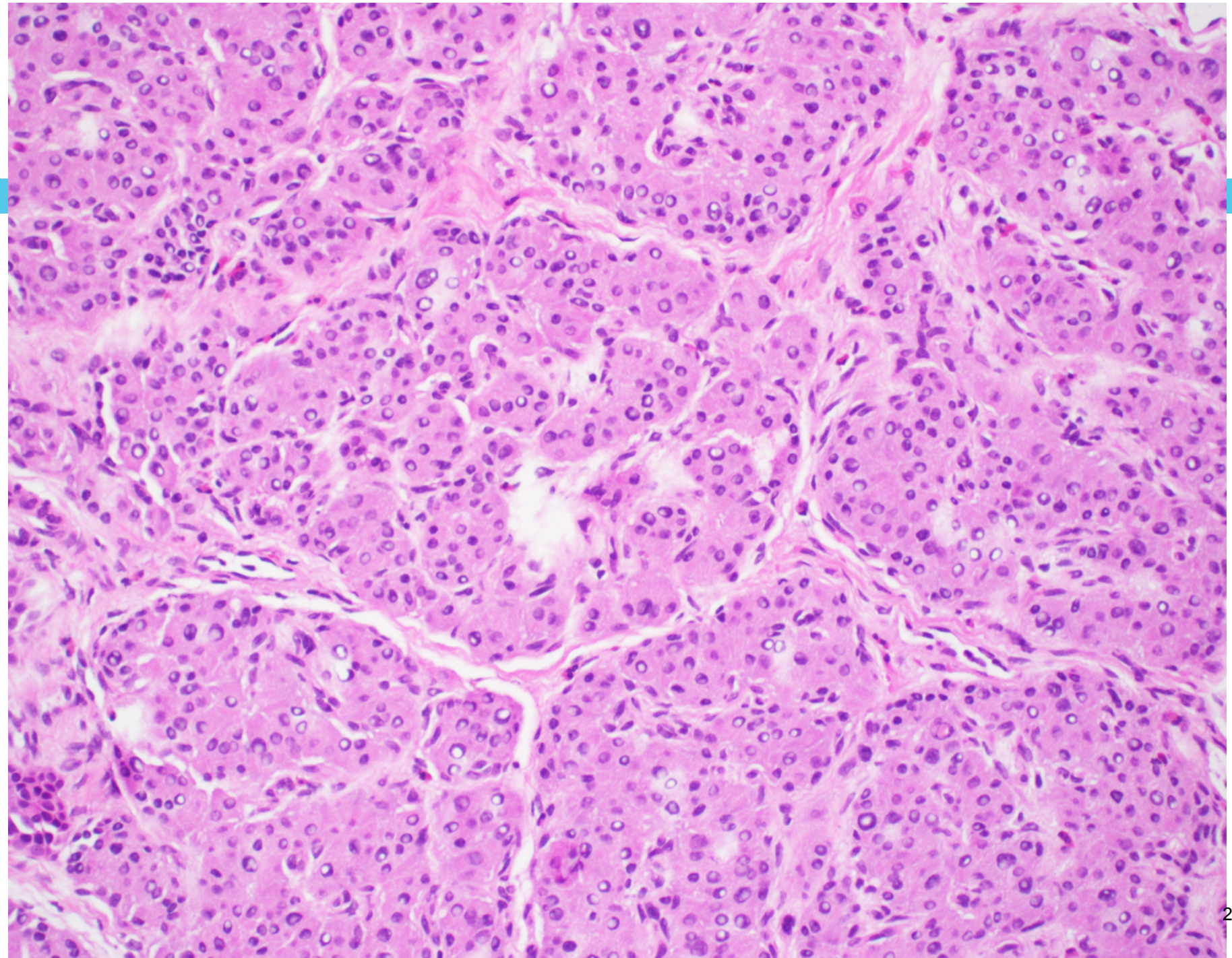

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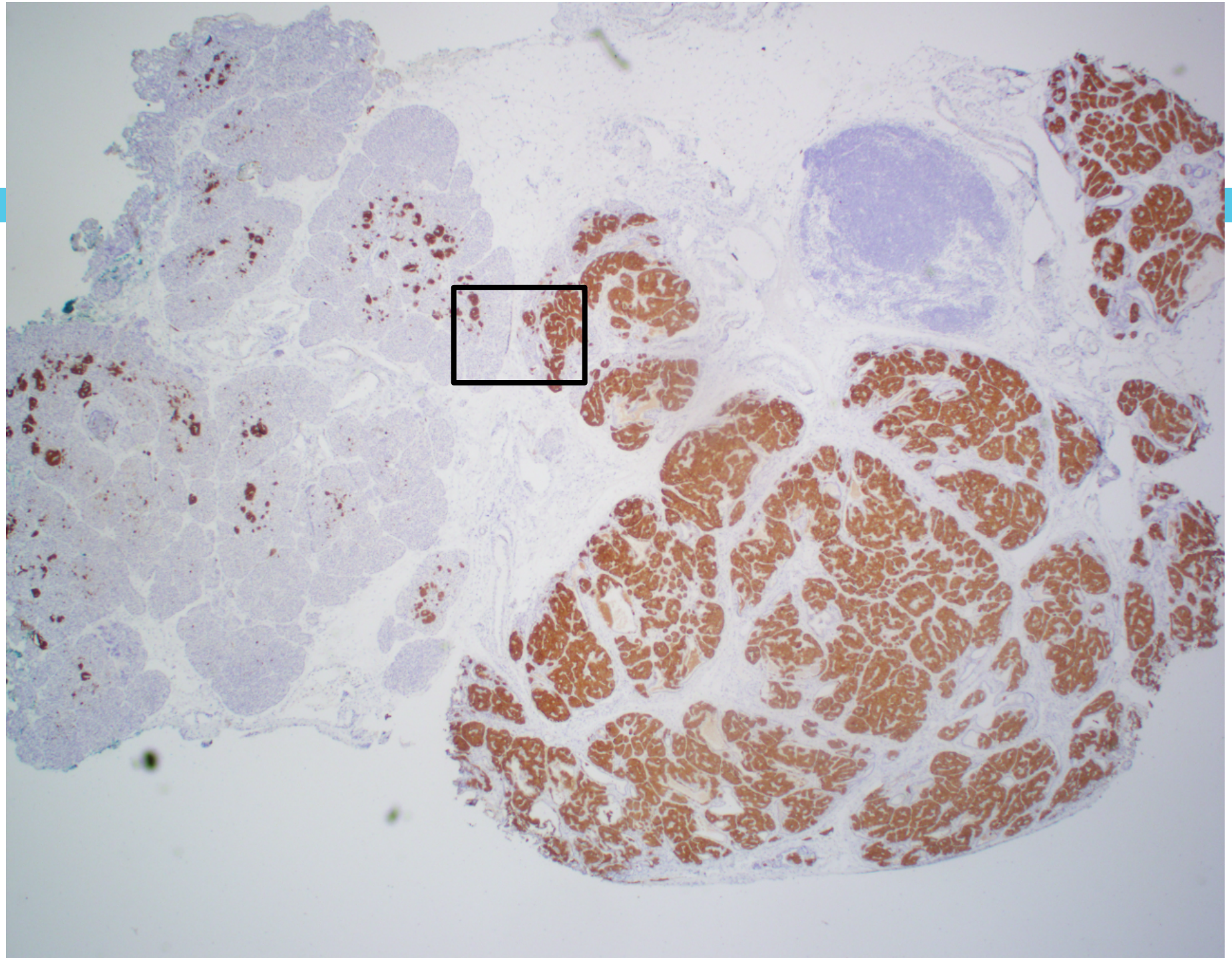

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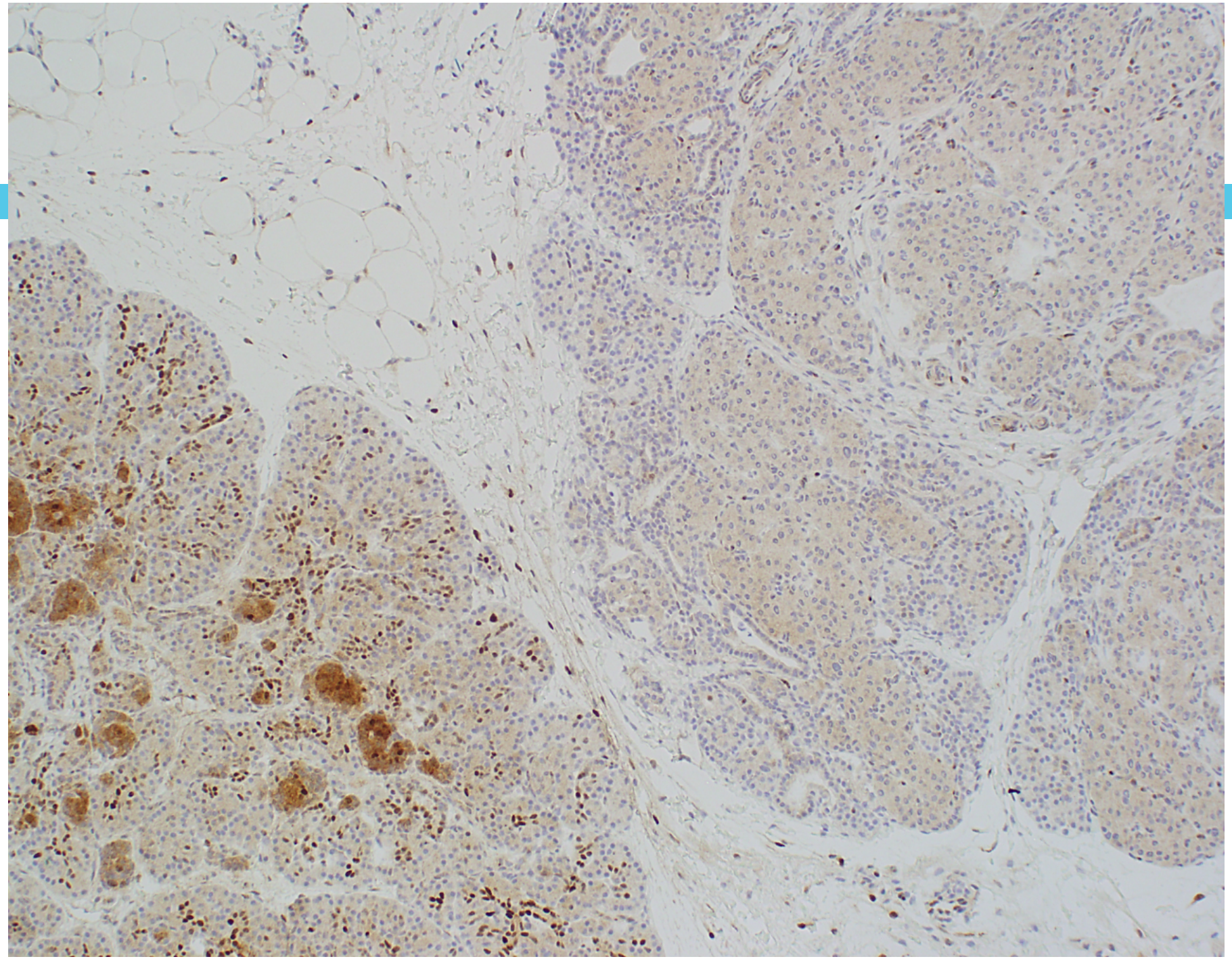
p57



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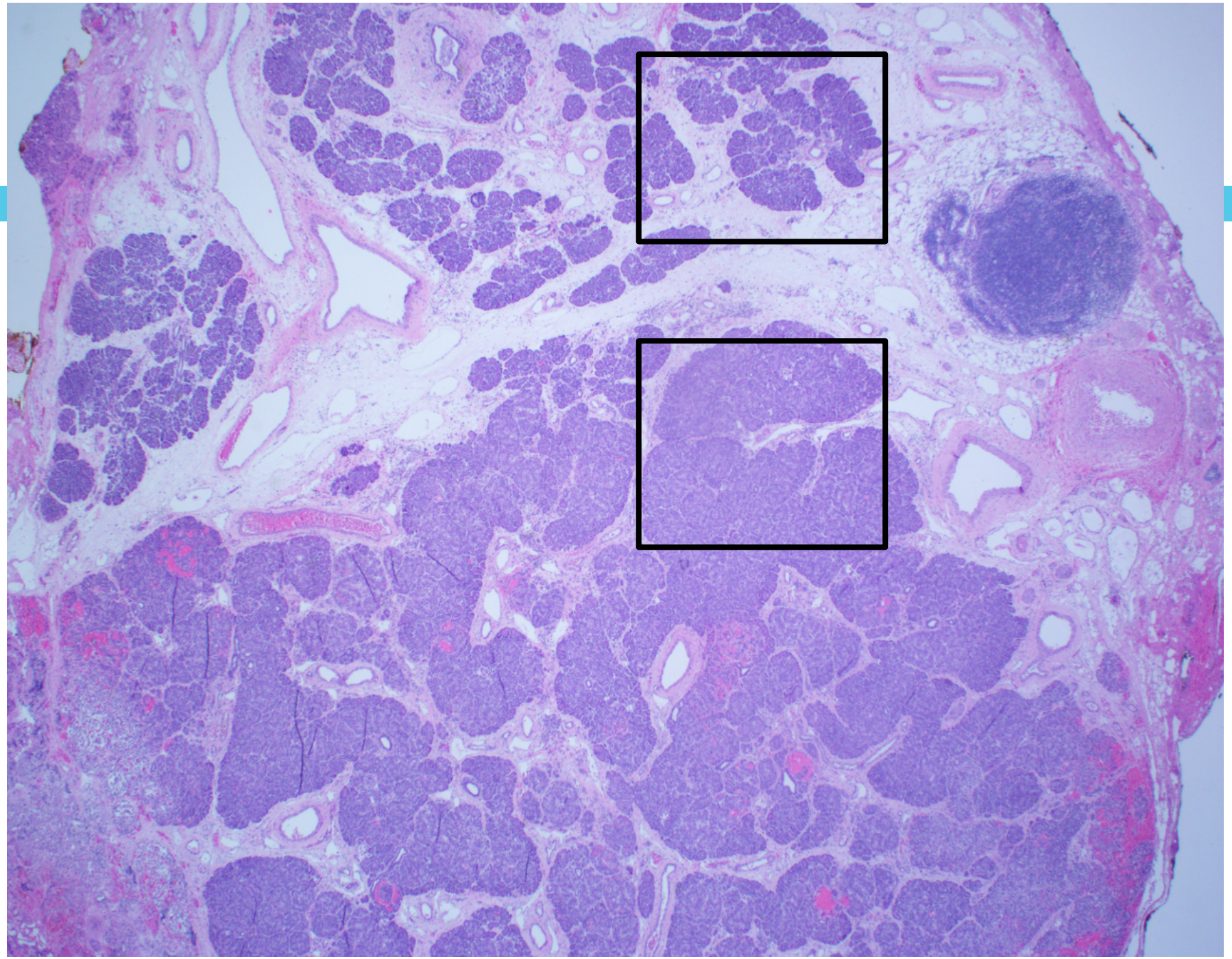
MIMIC OF FOCAL DISEASE (OVERGROWTH)



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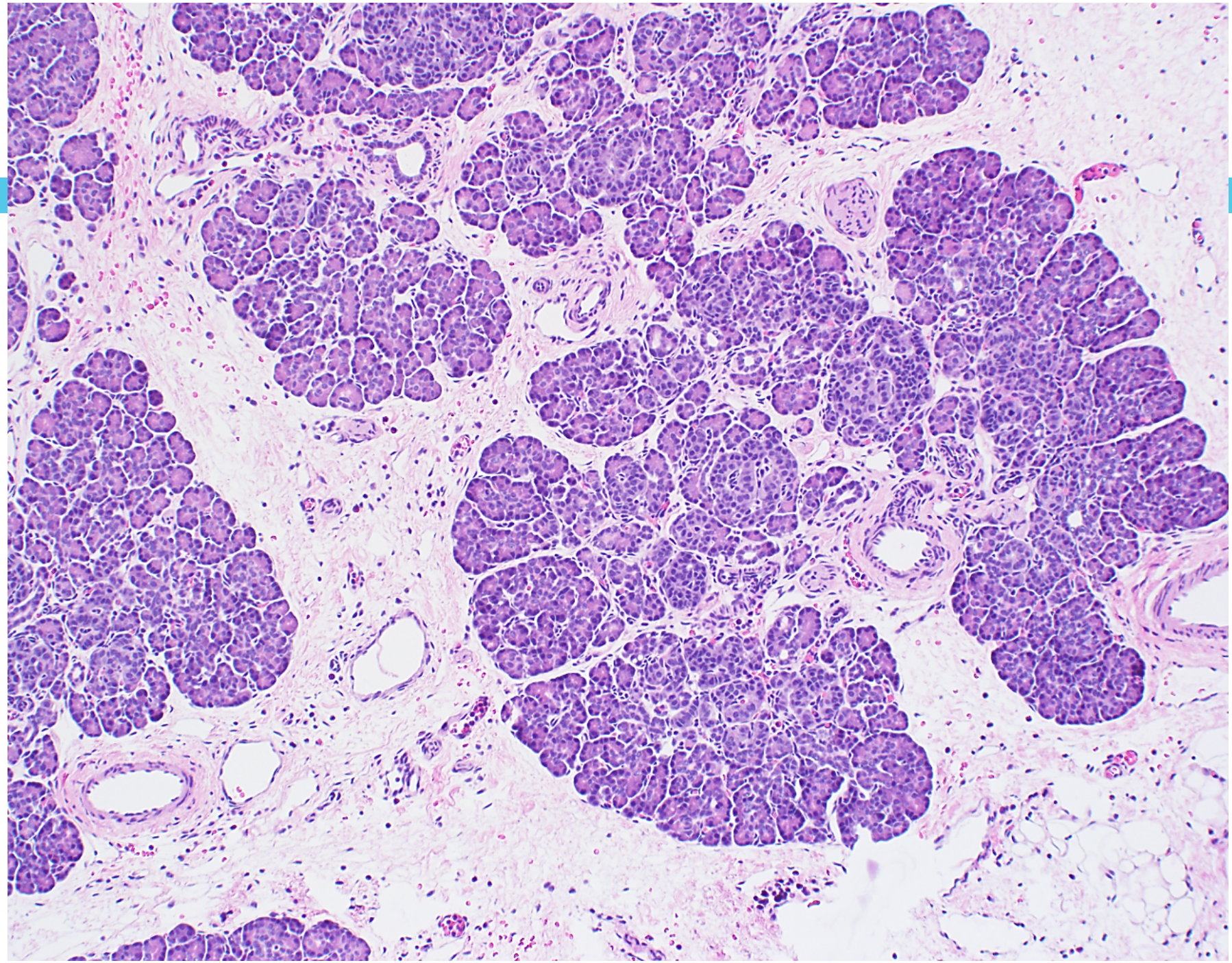

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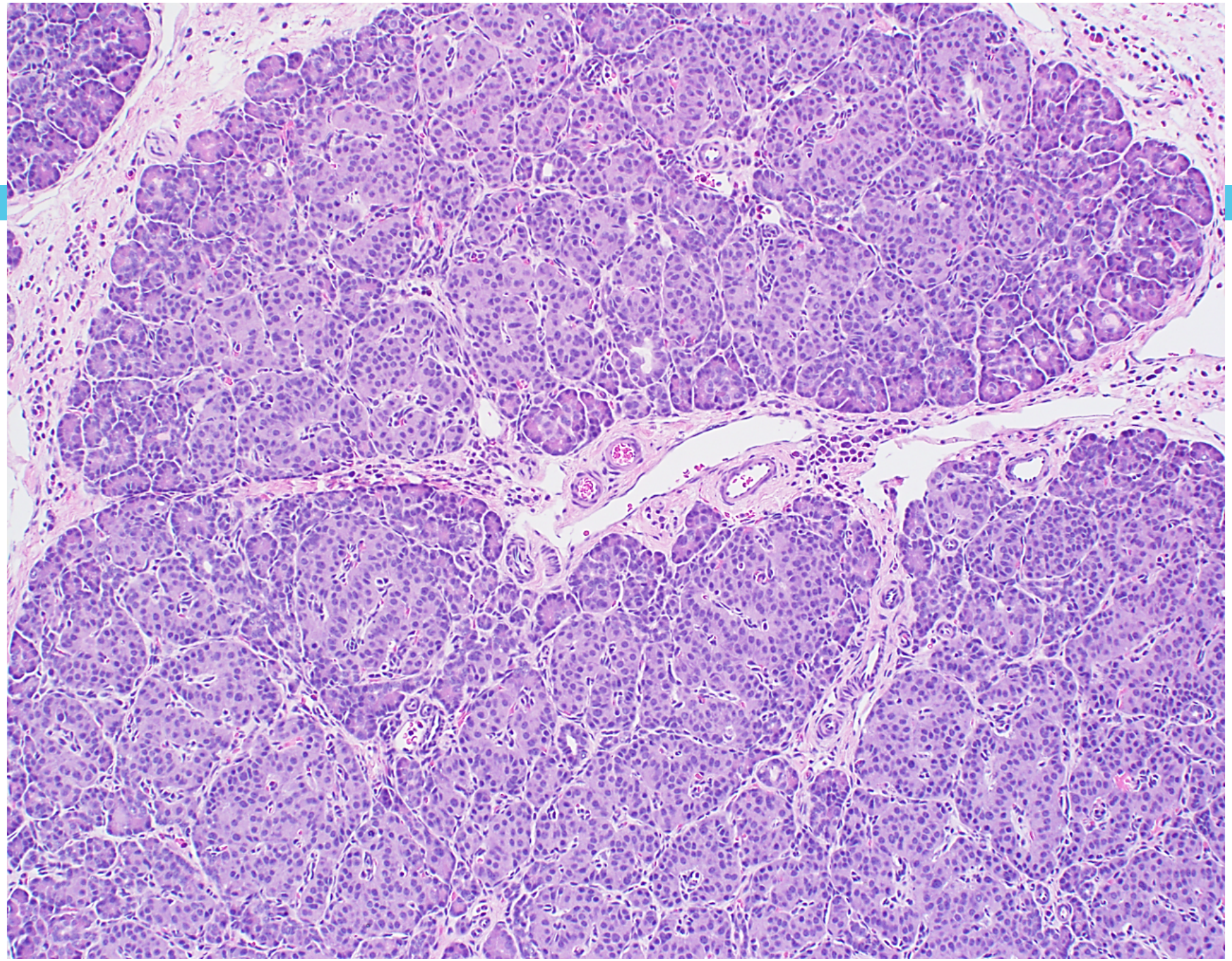

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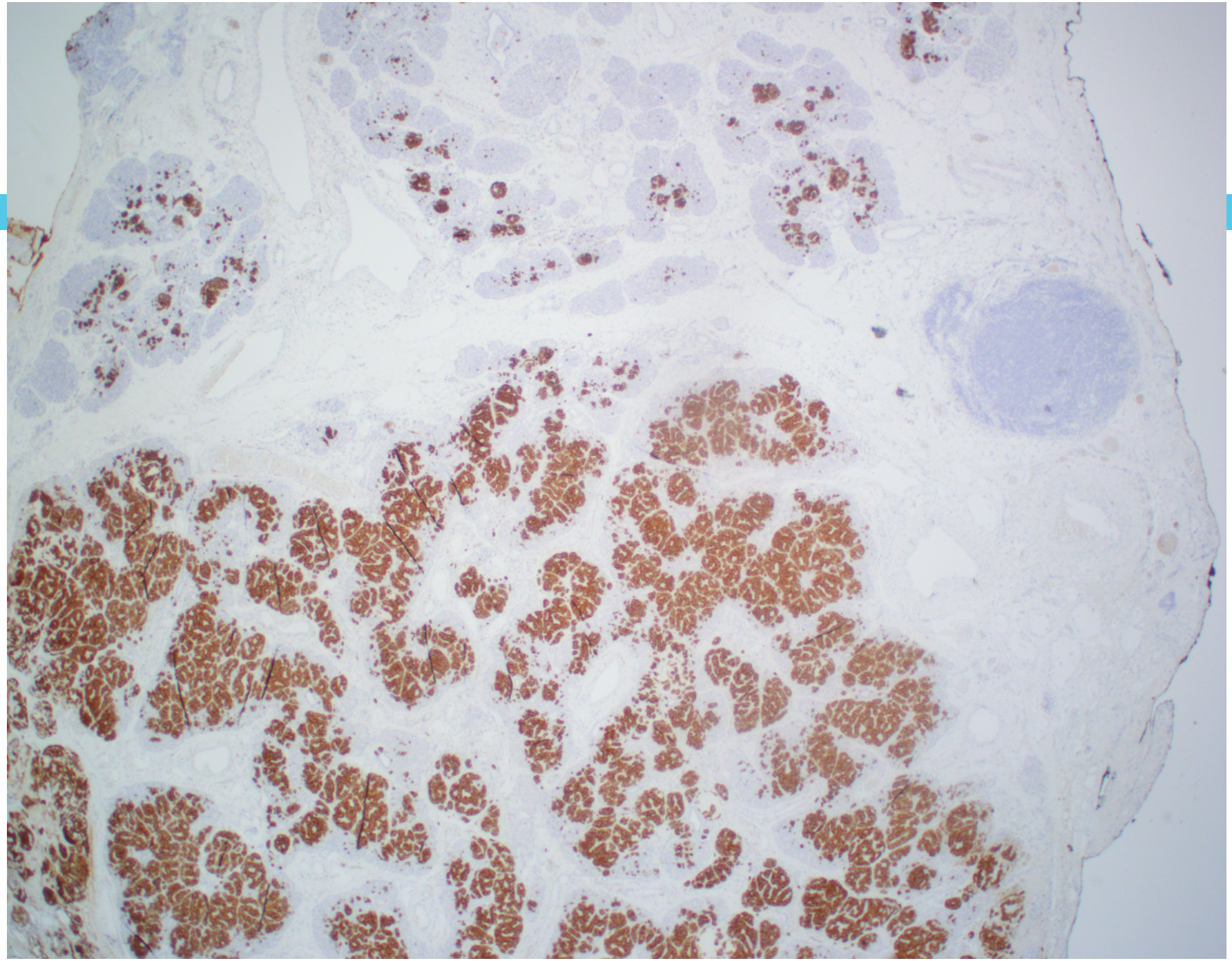

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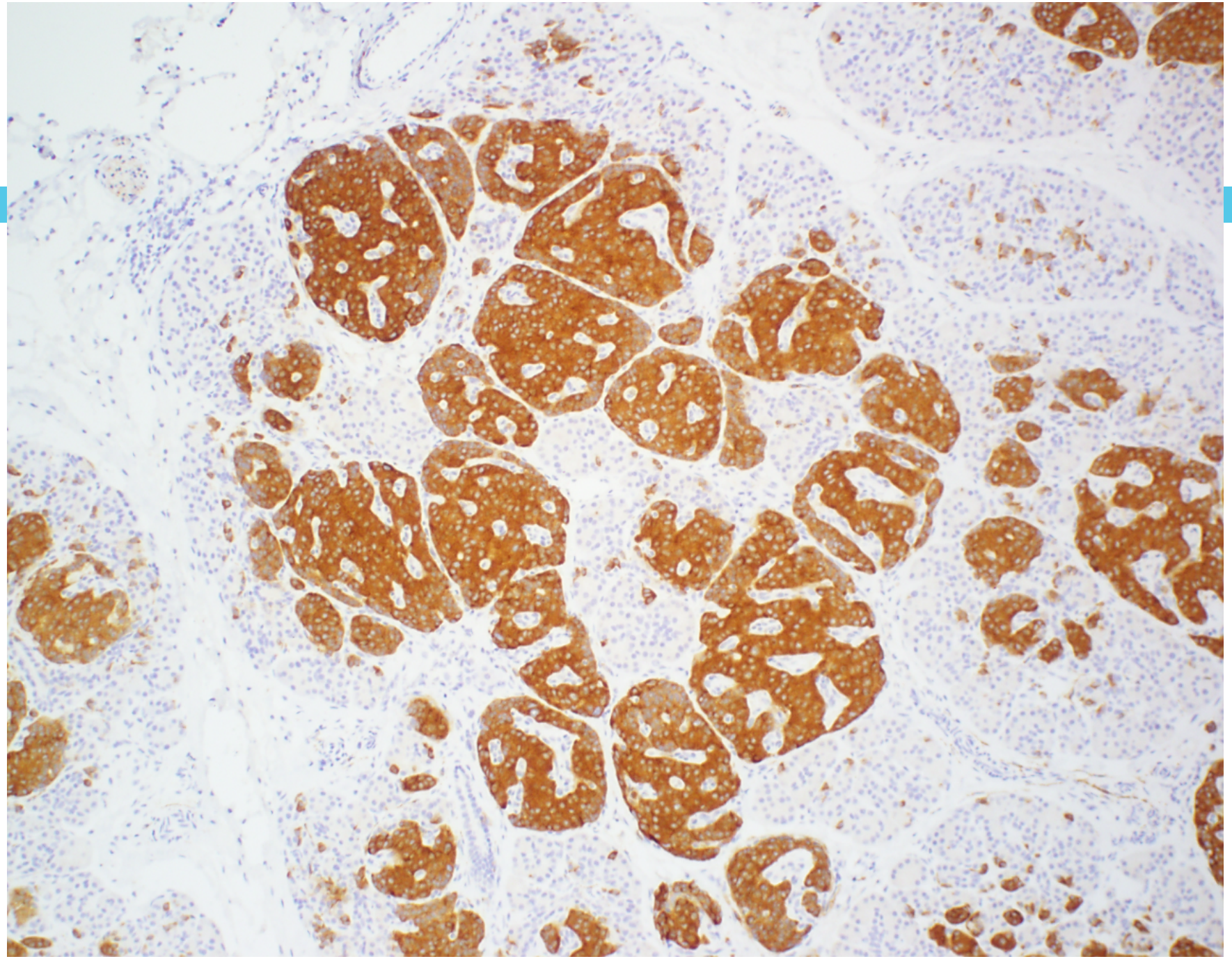

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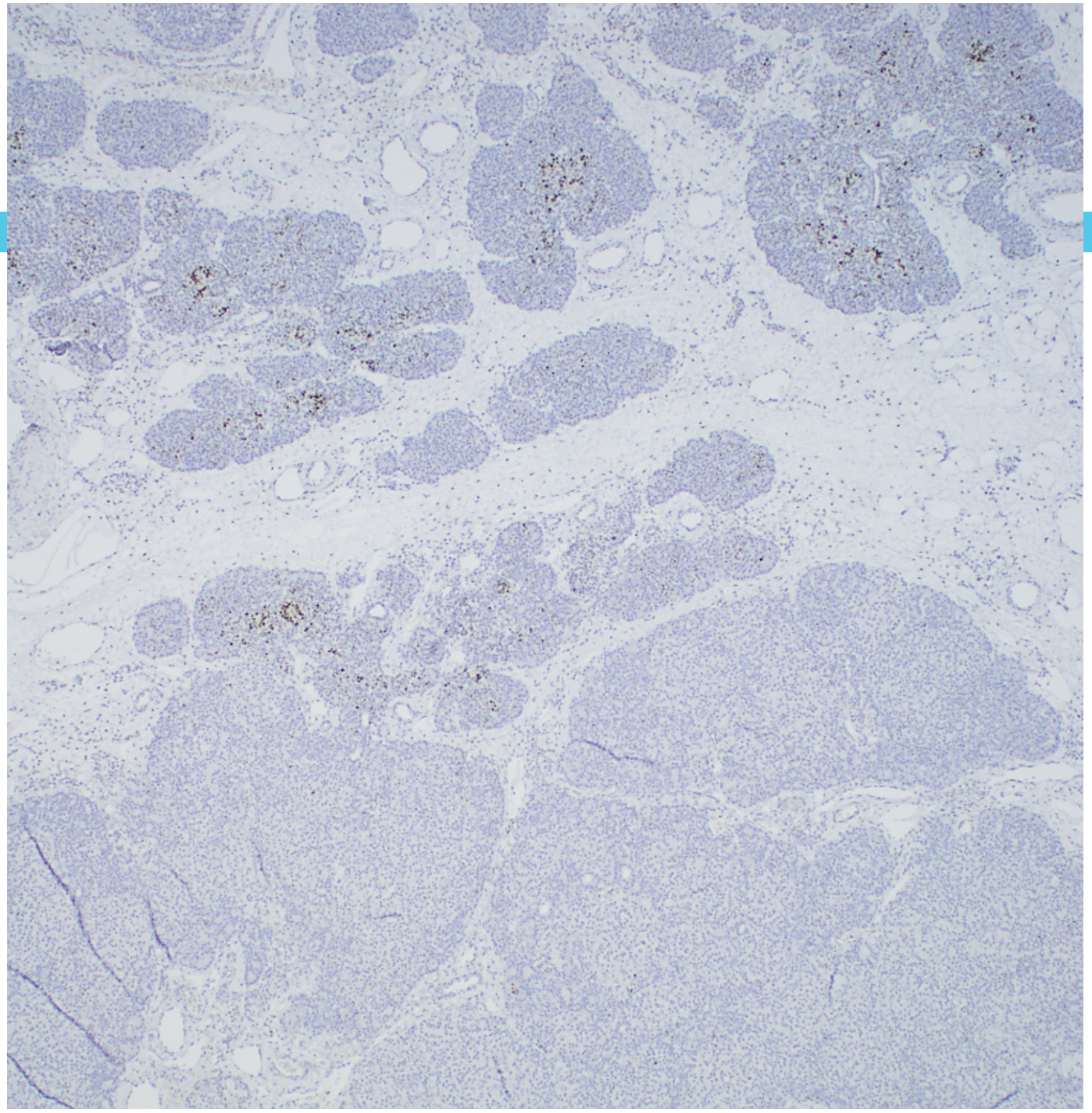
Loss of p57



Thank You!
Questions?

Contact
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