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Summary

NATURAL HISTORY OF PRSS1 HEREDITARY PANCREATITIS: A 30-YEAR FRENCH NATIONWIDE COHORT (PARADISIO-1)

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Gut Microbiota | Endoscopy | Radiology & Imaging | Pancreas

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Nationwide French cohort of 455 PRSS1 mutation carriers demonstrates 77.6% penetrance, median symptom onset at age 12.5 years, and a 32-fold increased risk of pancreatic cancer (SIR 32.2, 95% CI 19.4–50.3).

KEY POINTS

- 22 patients (4.8%) developed pancreatic ductal adenocarcinoma at a median age of 63.8 years, with diabetes independently associated with a 6.8-fold increased PDAC risk (95% CI 2.1–21.2).
- Pancreatic exocrine insufficiency occurred in 37.6% and diabetes mellitus in 21.5% of patients during follow-up.
- Median overall survival was 84.2 years, though overall mortality remained elevated compared to the general population (SMR 3.2, 95% CI 2.4–4.3).
- PRSS1 variants p.R122H and p.N29I were associated with earlier disease expression and increased risk of pancreatic insufficiency.
- The presenter recommended genetic testing in all relatives, lifelong clinical and biological follow-up even in asymptomatic patients, and continued pancreatic cancer surveillance.

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