



Cancer risk in severe alpha-1-antitrypsin deficiency

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Individuals with severe alpha-1-antitrypsin deficiency appear to have an increased risk of developing cancer compared with the general population, even after adjustment for age, sex, smoking habits and liver disease. <https://bit.ly/3ikSy0W>

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Abstract

Background Severe alpha-1-antitrypsin deficiency (AATD), phenotype PiZZ, is a risk factor for pulmonary emphysema and liver disease, but its effect on cancer risk is unknown. Our aim was to evaluate the risk and the risk factors for incident cancer in PiZZ individuals compared with the general population with known smoking habits.

Methods A longitudinal study of PiZZ individuals (n=1595) from the Swedish National AATD Register, and controls (n=5999) from Swedish population-based cohorts. Data on cancer and mortality were obtained by cross-linkage with national registers. Individuals who had undergone lung transplantation (n=10) and those with a cancer diagnosis within 5 years prior to inclusion (n=63) were excluded. The risk factors for developing cancer were analysed using proportional hazards and Fine–Gray regression models, adjusting for age, sex, smoking habits and the presence of liver disease.

Results The median follow-up time was 17 years (interquartile range 11 years) for the whole study population. The incidence rates of hepatic and non-hepatic cancer per 1000 person-years were 1.6 (95% CI 1.1–2.3) and 8.5 (95% CI 7.2–10.0), respectively, for the PiZZ individuals, and 0.1 (95% CI 0.04–0.2) and 6.6 (95% CI 6.0–7.1), respectively, for the controls. The adjusted hazard ratios for hepatic and for non-hepatic cancer were 23.4 (95% CI 9.9–55.4) and 1.3 (95% CI 1.1–1.5), respectively, in the PiZZ individuals compared with the controls.

Conclusion These results suggest that individuals with severe AATD may have an increased risk of developing both hepatic and non-hepatic cancer, compared with the general population.