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Title: Longitudinal follow-up of lung function in ataxia telangiectasia

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Body: Rationale: Individuals with Ataxia Telangiectasia (AT) are vulnerable to the development of significant pulmonary complications with age. This condition calls for a follow up on lung function evaluation. Objectives: To follow the individual patient's lung functions during 3-5 years Methods: Yearly best spirometry data were collected from 39 AT patients (age 10.4±5.3years; 13 patients younger than 7 years). The yearly decline in spirometry indices were defined in relation to the preceding year, initial age, and airway hyper-reactivity (a positive response to β₂-agonist). Results: Young children showed low FVC (%predicted) between 43.1-70.7%. Yearly decline in spirometry was -3.4±9.4%, -2.7±7.6%; -5.9±10.4; for FVC, FEV₁ and in PEF respectively. FEF₂₅₋₇₅ increased by 1.6±12.1%/year. Patients having airway hyper-reactivity (n=21) showed initially worse flows (%predicted) than others in FEV_{0.5} (41.6±18.5% vs. 57.7±15.7%, p<0.0055) and in FEF₂₅₋₇₅=83.3±19.2% vs. 106.3±20.8%; p<0.0006). Conclusions: Young A-T patients showed a restricted FVC pattern that deteriorated with age. Patients having hyperactive airways showed initially restrictive/obstructive worse lung function than others. Airway to volume ratio increases with age implying hypoventilation. The findings call for a restricted follow up on lung function in A-T patients.