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Title: Clinical progress over time in children with primary ciliary dyskinesia

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Body: Background Little is known regarding clinical progress over time in children with PCD, particularly once transferred to adult care. Aims The aim of this study was to look at clinical progress over a decade in a cohort of children with PCD first studied in 2002, measuring their lung function (% predicted FEV1), growth and chest exacerbations. Methods Ethical approval was obtained to approach subjects previously studied in 2002. Review of paper and electronic case notes of consenting subjects was conducted to determine hospital admission(s), lung function and nutritional parameters over time, and previous and current treatment. Results Eight out of 28 participants from the initial cohort were recruited into this study. Five participants in the initial study were not receiving any secondary care respiratory follow up and could not be contacted. There was a mean annual loss of 0.6% of % predicted FEV1, with % predicted FEV1 decreasing in 75% of participants. There was no statistical difference in the weight, height and BMI z scores at the two time points (current study period and initial study period in 2002) $p>0.05$. 37.5% of the participants had a gastrostomy in situ. Significantly one of the eight participants had undergone a bilateral lung transplant, and another had died with respiratory failure secondary to a respiratory tract infection at the age of 11. Conclusions Children with PCD can have significant respiratory disease resulting in death and lung transplantation. Follow-up regimens in adult respiratory clinics appear variable. There was no significant change in growth of individuals with PCD over time, however some participants were receiving nutritional support via a gastrostomy.