REFERENCES

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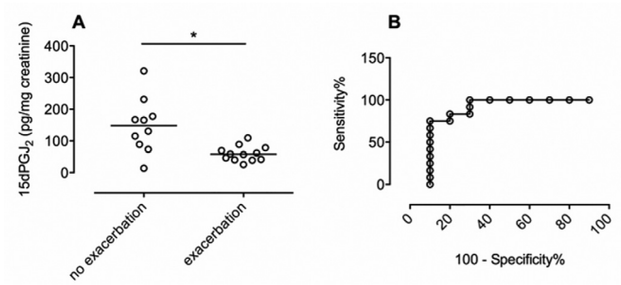

Abstract S65 Figure 1 (A) Urine 15dPGJ$_2$ at baseline is significantly lower in children who have an asthma exacerbation within 3 months. (B) ROC curve for 15dPGJ$_2$. ROC AUC=0.858, p=0.005. PG:prostaglandin. Bar represents median, comparison by Mann-Whitney test. *p<0.01.

S66 THE GLI SPIROMETRY REFERENCE EQUATIONS INFLUENCE THE APPARENT RATE OF DECLINE IN FEV1 AMONG CHILDREN AND ADOLESCENTS WITH CYSTIC FIBROSIS

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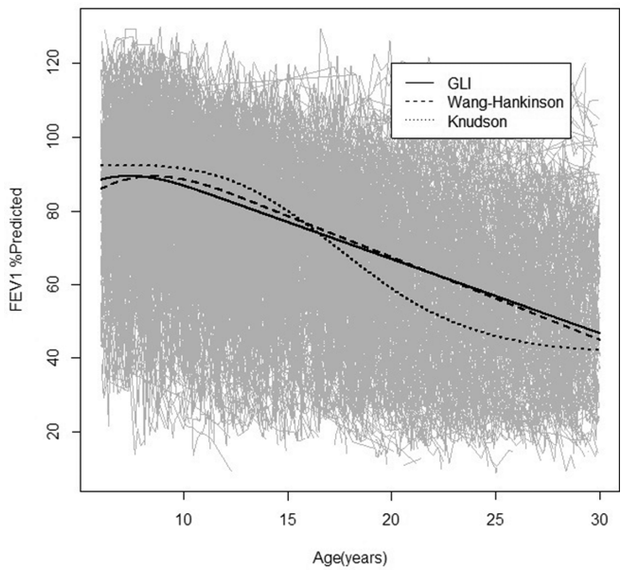
Background In patients with cystic fibrosis (CF), interpretation of cross-sectional FEV$_1$ data is greatly influenced by choice of spirometry reference equation, particularly during childhood (Stanojevic; J Cyst Fibros 2014). We hypothesised that availability of the Global Lung Function Initiative (GLI) spirometry reference equations (Quanjer; ERJ 2012) will also affect the apparent rate of decline in lung function over time, thereby potentially altering our understanding of disease progression in CF.



Abstract S66 Figure 1 Average FEV1 decline in people with cystic fibrosis according to three spirometry reference equations

Methods Data were extracted from two patient registries: the UK CF Registry (n = 6043 subjects; 20,013 test occasions over a period of 5 years) and the Toronto CF database (n = 1023 subjects; 27,868 test occasions over a period of 23 years). Spirometric outcomes were interpreted using%predicted FEV$_1$ calculated from GLI, Knudson (as currently used by the UK CF Registry), and Wang-Hankinson (as used by the US CF Foundation) reference equations. Patients >30 yrs or with FEV$_1$ > 130% predicted were excluded. We used a non-linear mixed effects model to describe the average change in FEV$_1$ with age. To illustrate the importance of reference equation in evaluating risk factors, FEV$_1$ decline according to patient gender was also explored.

Results The pattern of lung function decline at the population level differed according to selected equation (Figure). Average rate of decline was steeper with Knudson or Wang-Hankinson than GLI. Importantly, GLI equations showed a steady decline in FEV$_1$ starting at 6 yrs, whereas the other equations suggest greater decline during adolescence. Similar patterns were observed in both UK and Toronto populations. When analysed according to gender, the rate of lung function decline was steeper in females during early adolescence compared with males where the decline was steady.

Conclusions In both datasets, Knudson and Wang-Hankinson reference equations suggest relative preservation of spirometry in childhood followed by rapid decline in adolescence. However using the more robust GLI equations, steady decline throughout childhood with a less dramatic acceleration during adolescence is seen, with differences in pattern of change over time according to patient gender. Accurate identification of critical periods of lung function decline offers novel opportunities to target care.

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S67 LUNG CLEARANCE INDEX (LCI) IS A SENSITIVE PREDICTOR OF HIGH RESOLUTION COMPUTED TOMOGRAPHY (HRCT) SCORES IN CHILDREN WITH NON-CF BRONCHIECTASIS

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Introduction and objectives LCI is a sensitive predictor of early cystic fibrosis (CF) lung disease, and correlates with HRCT better than spirometry (Thorax. 2008;63:129–134). The same is true in adults with non-CF bronchiectasis (Am J Respir Crit Care Med. 2014;189:586–592.), but by contrast, in PCD there were no relationships between LCI, HRCT or spirometry (Am J Respir Crit Care Med. 2013;188:545–549). It is unclear whether these differences reflect primary versus secondary ciliary dyskinesia, or CFTR versus non-CFTR disease. We hypothesised that in children with non-CF bronchiectasis, relationships between spirometry, LCI and HRCT will be similar to those in CF children and non-CF bronchiectasis adults, rather than PCD patients.

Methods 12 children with non-CF bronchiectasis performed LCI and spirometry and underwent thoracic HRCT. HRCT scans were scored quantitatively (Thorax. 2013;68:532–539). Results were compared with those from large CF (n = 125) and PCD (n = 38) cohorts.

Results In non-CF bronchiectasis there was a correlation between first second forced expired volume (FEV$_1$) and LCI (p = 0.009, r=-0.6), similar to that seen in CF (p < 0.0001,