

the patient admitted to any problems at all while driving (P=0.0098, OR=1.6). Males were more likely to allow OSAS patients to drive. Consultants without an interest in sleep medicine and respondents seeing less than 5 patients per month were more likely to advise against driving. The advice given was not related to the age of the clinician.

**Conclusions** This survey has shown that there is considerable variability in the advice given by clinicians, with in some situations a patient having an even chance of receiving opposite advice, depending on who they see. Restriction of driving has major implications for an individual, both social and financial. Allowing someone to drive who is not safe to do so has potentially disastrous consequences for them and others. The issue of how to improve consistency of advice needs to be addressed.

## Non-invasive markers of lung disease in children

### S6 LONG TERM LUNG FUNCTION OUTCOMES IN CHILDREN BORN WITH ABDOMINAL WALL DEFECTS

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**Introduction** Although survival following surgical repair of abdominal wall defect is now good, there are very few data regarding long-term lung function (LF) outcomes. The aim of this study was to describe LF at school-age for a post-surgery population from a single large centre.

**Methods** All children having surgical repair of exomphalos or gastroschisis between 1/1/1998–30/6/2007 at our institution were identified, and those aged greater than 6 years were recalled for LF testing. Children born with major associated cardiac, genetic or laryngeal conditions were excluded from the study. All children underwent resting oximetry, body plethysmography, forced spirometry and carbon monoxide transfer test. Clinical and demographic data was also collected.

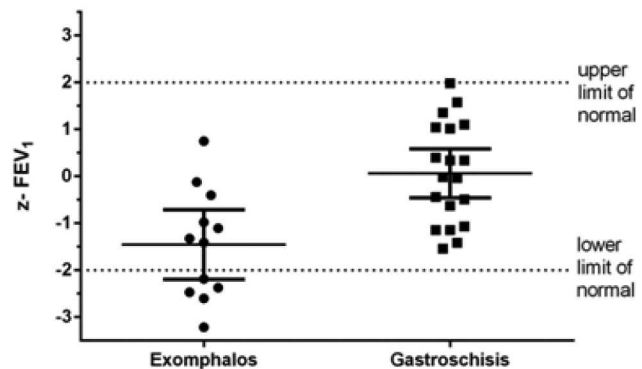
Results were converted to z-scores to adjust for height, sex, age and ethnicity. Statistical comparisons between groups were performed with t-tests.

**Results** 86 children were approached and LF data were obtained in 31 (12 Exomphalos;19 Gastroschisis) children aged  $\geq 6$  years. 22 children were male; the mean age at testing was 9.9 years (range: 6.4–14.4). When analysed as a group these children had significantly reduced forced expiratory volumes in 1s, FEV<sub>1</sub> ( -0.52 z-scores [95%CI -1.01,-0.04] p = 0.036), FEV<sub>1</sub>/FVC (-0.88 [-1.21, -0.56] p < 0.000), lung transfer factor, TL<sub>CO</sub>(-0.76 [-1.05,-0.47] p < 0.000), TL<sub>CO</sub>/VA (-1.77 [-2.08,-1.46] p < 0.000), functional residual capacity, FRC (-0.45 [-0.70, -0.19] p = 0.001), residual volume, RV (-0.36 [-0.61,-0.12] p = 0.006) and significantly increased alveolar volume, VA (0.95 [0.37, 1.53] p = 0.002). Forced vital capacity, FVC was normal (-0.01 [-0.54, 0.51] p = 0.96)

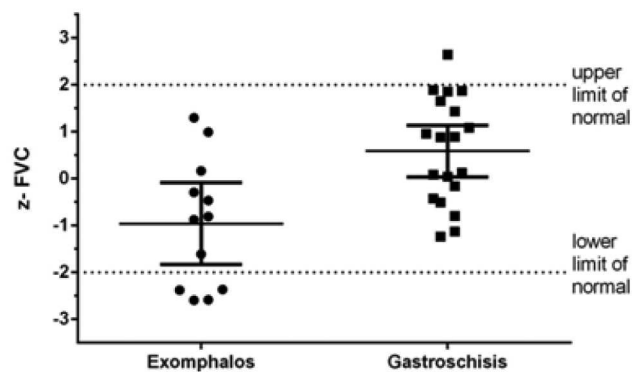
When analysed as subgroups, children post exomphalos repair had significantly lower LF than those post gastroschisis repair, with a mean difference (95% CI of difference [Exomphalos–Gastroschisis] ) in FEV<sub>1</sub> of -1.52 z-scores (-2.35, -0.68; p = 0.001); FVC -1.54 (-2.47, -0.62; p = 0.002); TLC -0.91 (-1.78, -0.04; p = 0.041). No significant group differences were found in the remaining lung function outcomes.

**Conclusions** This is the largest study to report lung function at school-age in children post abdominal wall surgery. We demonstrate that these children have significantly reduced lung function; and when analysed by subgroup, that those with a history of exomphalos repair have significantly greater defect than those with a history of gastroschisis repair.

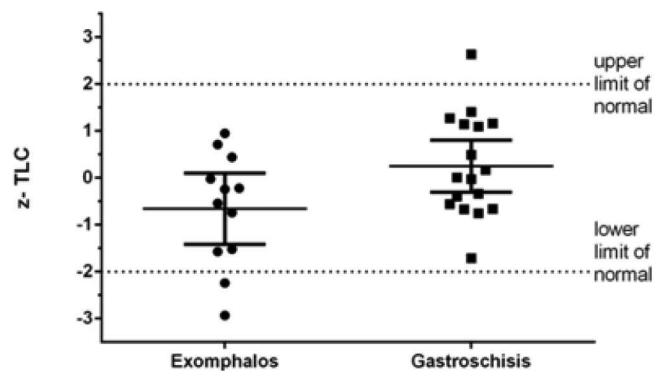
#### Comparison of Forced Expiratory Volumes in 1 second



#### Comparison of Forced Vital Capacity



#### Comparison of Total Lung Capacity



**Abstract S6 Figure 1. Comparison of lung function outcomes between groups. Spirometry and plethysmography z-scores derived from Global Lung Function Initiative 2012 and Rosenthal 1993 equations respectively.**

### S7 EVOLUTION OF LUNG FUNCTION DURING THE FIRST TWO YEARS OF LIFE IN INFANTS WITH CYSTIC FIBROSIS DIAGNOSED BY NEWBORN SCREENING

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**Background** In contrast to findings from the Australian AREST-CF study<sup>1</sup> infants with cystic fibrosis (CF) diagnosed by newborn screening (NBS) participating in the London CF Collaboration (LCFC) study<sup>2</sup> were found to have relatively mild lung disease by 1 year of age when compared to healthy controls.

**Hypothesis** In NBS CF infants, lung function remains stable to 2 years.

**Methods** Lung clearance index (LCI), plethysmographic functional residual capacity (FRC) and forced expiratory volume (FEV<sub>0.5</sub>) from the raised volume technique were measured in NBS CF infants and healthy controls at 3 months (3m), 1 year (1y) and 2 years (2y). Results were expressed as z-scores using equipment-specific equations to adjust for age, sex and body size.

**Results** To date, 55 CF and 28 control infants have been assessed on all 3 occasions. The mildly elevated LCI and FRC in

CF infants identified by 3m when compared with controls remained stable thereafter. The significant reduction in FEV<sub>0.5</sub> (mean difference (95% CI) -1.26 (-1.73; -0.79) z score) among CF infants at 3m had improved by 1y (see Figure 1). From 1 to 2 years all 3 measurements remained stable with no significant changes in average z-scores for either the CF or control infants. On average, LCI, FRC and FEV<sub>0.5</sub> only changed by 0.02, 0.16 and 0.06 z-scores respectively amongst CF children between 1–2 years, similar to that observed in controls. Mean (95%CI) group differences (CF-HC) in change of LCI, FRC and FEV<sub>0.5</sub> between 1–2y were 0.02 (-0.61; 0.66), p = 0.94; -0.05 (-0.61; 0.51), p = 0.86; and 0.32 (-0.27; 0.90) p = 0.29, respectively.

**Conclusions** This is the first study to demonstrate stable lung function to 2y in NBS CF infants managed on standard CF therapy. These results suggest that in many of these infants novel treatments could be deferred beyond infancy when objective outcomes are more easily measured.

## REFERENCES

1. Pillarisetti *et al*. *AJRCCM* 2011
2. Hoo *et al* *Thorax* 2012.

## S8 HYPERPOLARISED 3HE MRI IS SUPERIOR TO LUNG CLEARANCE INDEX IN DETECTION OF VENTILATION ABNORMALITIES IN YOUNG CHILDREN WITH MILD CF

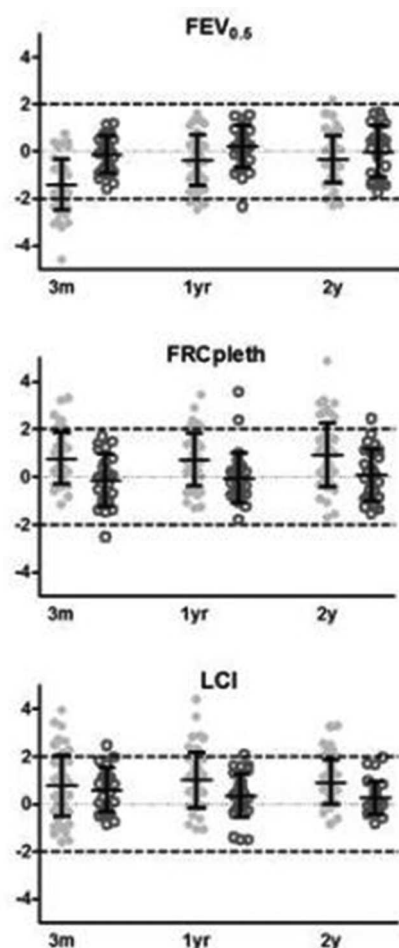
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**Introduction** Hyperpolarised <sup>3</sup>He MRI provides high resolution images of lung ventilation and is more sensitive than spirometry to early changes in lung ventilation in cystic fibrosis (CF). Lung clearance index (LCI) is a global measure of ventilation heterogeneity which is also sensitive to early changes in the lungs in CF before spirometry. The aim of this study was to investigate the capability of hyperpolarised <sup>3</sup>He MRI and LCI to detect ventilation changes in children with mild CF.

**Methods** 4 CF patients (FEV<sub>1</sub> 78–110% predicted) and 4 healthy volunteers have been assessed so far. <sup>3</sup>He ventilation images were acquired at breath-hold following inhalation of hyperpolarised <sup>3</sup>He, with 2.7x2.7x10mm resolution and full lung coverage using a 1.5T MRI system. The percentage of lung ventilated (VV%) was calculated as <sup>3</sup>He ventilated volume divided by total lung volume segmented from <sup>1</sup>H MR images. LCI was measured using 0.2% SF<sub>6</sub> and a modified Innocor gas analyser. LCI was performed sitting and repeated supine to mimic the position adopted for MRI scanning. Spirometry and plethysmography were also performed. Gas trapping was calculated as % difference in plethysmographic versus washout FRC.

**Results** Healthy volunteers had a mean (standard deviation) age of 8.8(1.5) years, FEV<sub>1</sub>% predicted = 97(10) and gas trapping = 4.4(9.1)%. LCI sitting was 6.9,7.3,6.6 and 6.9, LCI supine was 7.3,7.5,6.8 and 6.4. Healthy volunteers had homogeneous ventilation in <sup>3</sup>He ventilation images e.g. Fig1(a), and VV% = 94.5(2.8). CF patients had an age of 11.8(2.9) years, FEV<sub>1</sub>% predicted = 95(13), and gas trapping = 8.7(11.0). LCI sitting was 7.7,6.6,6.6 and 9.1, LCI supine was 7.8,7.3,7.1 and 11.8. Ventilation abnormalities were observed using <sup>3</sup>He MRI in all four CF patients scanned (Fig 1(b-e), with order



**Abstract S7 Figure 1.** Comparison of change in lung function z-scores between 3m, 1y and 2y in NBS CF infants and healthy controls. Legend: Data are expressed as mean (SD). Closed circles represent NBS CF infants; open circles represent healthy controls. Lung function outcomes were expressed as z-scores which adjusted for age and body size as appropriate. Dashed lines indicate limits of normality (+/- 2 z-scores).