479. New understanding of childhood lung disease through physiological measurement

P4598
Plp1 mutation induces altered respiratory response to an airway challenge
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Pelizaeus-Merzbacher disease (PMD) is a disease caused by mutations of the proteolipid protein1 (PLP1) gene that result in defective CNS myelination. Mice with an extra copy of Plp1, called Plp1/dup, develop a syndrome that models the duplication form of PMD. Patients with all except the mildest forms have respiratory involvement.

Objective: We hypothesized that Plp1/dup mice would lack protective airway responsiveness (AR) to an autonomic drug challenge. To address this, we investigated whether respiratory mechanics in these mice would be different at baseline (BL) or during methacholine (MCh) challenge.

Methods: Wild type (WT) n=16, carrier (Car) n=8 & affected (Af) n=17 mice, 3 months (3m) and 6 months old (6m), were anesthetized, mechanically ventilated & challenged with 0.1-6 mg/ml of aerosolized MCh. We calculated resistance (R), dynamic/static compliance (Cdyn/Cstat), asynchrony (PhRTB); lung tissue biomarkers & histological analysis are ongoing.

Results: BL differences were found in R, between WT vs Car (p<0.05) dependent on group (p<0.001) & age (p<0.0001), and in Cdyn, among WT vs. AF (p<0.01). MCh increased R as a function of dose in WT & Car, whereas Af6 mice lacked sensitivity to MCh (p<0.05). No differences in body weight, gender (WT females vs. WT Males) and Cotat were found. Af6 mice had the highest PhRTB.

Conclusions: WT mice compared with Af mice were lacking of AR to MCh at 6m, but not at 3m. These results indicate an age-associated lack of protective autonomic AR in the Plp1/dup mouse model and suggest that respiratory autonomic disequilibrium may contribute to the respiratory involvement in PMD patients. The Plp1/dup animal model may be used for testing therapeutic interventions.

P4599
Bronchiolar wall structure is altered in adult mice following neonatal hyperoxia
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Background: Very preterm infants often require supplemental oxygen, and can have an increased risk of poor lung function and asthma in later life. Follow-up studies suggest that factors associated with very prematurity can contribute to the development of chronic lung disease in the small conducting airways.

Objective: To determine if bronchiolar wall structure is persistently affected by neonatal exposure to hyperoxic gas.

Methods: Neonatal mice (C57BL/6J) breathed 65% O2 from birth until postnatal day 7 (P7), after which they lived in room air until P56 (n=26). Controls breathed room air from birth (n=25). Bronchiolar walls were histologically assessed for thickness, proportions of proliferating epithelial cells, ciliated and Clara cells, the amount of collagen and airway smooth muscle (ASM). The results were compared using one-way ANOVA.

Results: AF6 mice lacked sensitivity to MCh (p<0.05) compared to controls. MCh increased R as a function of dose in Wt6 & Car6, whereas Af6 mice had the highest PhRTB.

Conclusions: AF mice compared with WT mice were lacking of AR to MCh at 6m, but not at 3m. These results indicate an age-associated lack of protective autonomic AR in the Plp1/dup mouse model and suggest that respiratory autonomic disequilibrium may contribute to the respiratory involvement in PMD patients. The Plp1/dup animal model may be used for testing therapeutic interventions.
**P4601**

Routeline measurement of the LCI in CF with an ultrasonic device for multiple 
breakthrough nitrogren washout

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During the last decade multiple breath washout technique (MBW) for calculating the Lung Clearance Index (LCI) has become very popular for assessing ventilation inhomogeneity (VI) as an early manifestation of chronic lung disease. However, routine use has been difficult not only due to limited availability of licensed equipment. Inert tracer gases (e.g. SF6, He) certified for medical purporses are not universally available. Switch to nitrogen washout (MBW N2) using 100% oxygen may overcome this problem.

The aim of this cross sectional study was to assess whether LCI derived from MBW N2 discriminates as well as MBW SF6 between patients with CF and healthy controls. 19 controls (7-15 years) and 11 unselected patients with CF (7-25 years) performed three 2-3 minute MBW tests using the using the EasyOne Pro LAB™ (ND Switzerland) with 100% oxygen.

Mean (SD) LCI was 6.5 (0.64) in controls and 9.3 (1.93) in CF with a mean difference (95% CI, p-value) of -2.83 (-4.14-1.51, 0.001) between the groups. Within-test repeatability (CV%) was 5.3% in controls and 7.7% in CF.

Assessment of LCI using licensed equipment for MBW SF6 was feasible and well tolerated in both. In children and adults patients and controls. LCI based on MBW SF6 differed significantly between patients with CF and controls and results were comparable to published data obtained with different equipment and with using SF6 as tracer gas. We conclude that MBW N2 reflects VI similar to MBW SF6 and may thus be used for clinical application of MBW in patients with CF.

**P4602**

Bench test of a mass spectrometer based multiple-breath washout system using a realistic lung model

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**Background:** Lung volume measurement accuracy of multiple-breath washout (MBW) systems should be validated under realistic conditions. Here we used a lung model previously shown to be suitable for MBW system validation under physiological conditions, the custom mass spectrometer MBW system previously described for clinical use in adult and pediatric populations.

**Methods:** Functional residual capacity (FRC) measurement accuracy was assessed across a range of FRCs (430-4120 mL), tidal volumes (210-1010 mL), and respiratory rates (36-12 min-1). A wash-in inert gas mixture containing 4% sulfur hexafluoride (SF6) and 4% helium (He) was used for MBW. 63 MBWs were conducted over two test days. Measured gas and flow signals were processed in custom software (TestPoint). FRC was calculated as cumulative inert gas expired volume divided by the difference of MBW start minus end inert gas concentration.

**Results:** Mean (range) coefficient of variation of FRC was 0.6% (0.1-2.2%). Mean difference between measured and nominal FRC was 46.7 mL (1.7%) and lower limits of agreement were 155.3 mL (5.1%) and -62.0 mL (-1.8%), respectively. Of 126 FRCs, 124 (98%) were within 5% of the nominal FRC, mean error (SD) was -0.7% (0.4%). Error was associated with respiratory rate (R2 = 0.23).

**Conclusion:** Using a lung model previously shown to be suitable for MBW system validation under physiological conditions, the custom mass spectrometer MBW system accurately and reproducibly measures FRC’s ranging from preschool to adult lung volumes.

**P4603**

Validation of multiple breath washout technology in healthy children and children with CF

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**Mass spectrometry based technology is considered the “gold standard” for measuring LC, but is not readily available. In this ongoing study we aim to validate a nitrogen washout system for use in CF children. In a cross-over design, healthy and healthy children and CF children performed MBW by mass spectrometry (AMIS 2000; Innovision A/S, Odense, Denmark) using 4% SF6 or by nitrogen washout (Exhalalyzer D, Eco Medics AG, Switzerland). Results were independently scored by two operators; Bland-Altman plots were used to assess the agreement between the two tests. To date 24 healthy children (median age 11 years (range 3-17)) and 33 children with CF (median age 11 years (range 3-17)) completed MBW measurements using both the mass spectrometry and N2 washout. There was no systematic bias observed in LCI between the two methods. Overall there was good agreement healthy children (95% of all measurements agreed within -0.44; 0.83); however LCI N2 was systematically 0.2% (0.96; 0.33) units lower than LCI SF6.**

**P4604**

Ways to shorten the lung clearance index measurement LCI – Are there any measurements needed?

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**Background:** Inert gas multiple-breath washout (MBW) derived lung clearance index (LCI) is a sensitive lung function parameter in subjects with mild cystic fibrosis (CF) lung disease, but rarely measured in clinical routine due to lack of available equipment and lengthy protocols. Using an available nitrogen (N2) MBW setup (Exhalalyzer D, Eco Medics, Switzerland) we assessed shortened N2 MBW protocols for LCI.

**Methods:** We determined whether the LCI from the 1st (LCI1) and the mean LCI of the 1st and 2nd valid N2 MBW (LCI2), respectively, are comparable to the averaged information obtained from three N2 MBW (LCI3).

**Results:** LCI1, LCI2, and LCI3 differed significantly between healthy and CF children. LCI1 and LCI2 were strongly associated with LCI3 (R2 = 0.98 for both), took less time, and were of similar diagnostic value. Comparing LCI1 and LCI2, mean (range) test duration was 2.2 (0.5-5.2), 6.6 (3.6-12.7), and 11.0 (8.0-17.1) min, and upper limits of normal LCI (8.3, 8.3, and 8.4) all correctly classified 71% of children, respectively. Bland-Altman analysis of LCI1 and LCI2 showed good agreement with LCI3: Mean difference was -1.1% and -0.7%, limits of agreement were 6.8 to -9.0% and 5.9 to -7.3%, respectively.

**Conclusion:** In the current study population, the 1st LCI or the mean of the 1st and 2nd LCI take less time and predict the mean LCI of three N2 MBW within physiological measurement variability. Using less N2 MBW measurements seems promising for time-saving LCI measurement in clinical routine.

**P4605**

Abnormalities in lung clearance index in CF infants diagnosed by newborn screening

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**Newborn screening (NBS) in CF offers the potential to prevent lung damage before the onset of clinical symptoms. The lung clearance index (LCI), measured by multiple breath washout has shown promise as a marker of early lung disease in patients with CF. Therefore, we aimed to determine whether LCI could be used to distinguish between subjects with CF and healthy subjects in the first 2 years of life. Healthy infants (3 months – 2 years) completed MBW testing as part of the infant pulmonary function protocol of the Canadian Healthy Infant Longitudinal Development Study (CHILD Study). Infants (3 months – 2 years) with diagnosed CF attending the Respiratory Medicine Clinic at the Hospital for Sick Children were invited to complete MBW testing. MBW was measured 51 healthy infants and 18 infants with CF diagnosed by NBS. Despite NBS, infants born with CF were smaller (Height-for-age z-score (-1.19 (-0.58; -1.81) and lighter (mean difference weight-for-age z-score (-0.40 (-0.98; 0.19) compared with healthy controls. The LCI was on average 0.4 units higher in CF (median LCI 7.32 (SD 0.91)) compared with healthy infants (6.92 (SD 0.61)). Adjusting for the relationship between LCI and height in the first 2 years of life, LCI was 0.67 (95% CI -1.34; 0.01) z-scores higher in CF compared to healthy infants, although overlap was considerable (Figure). Therefore, despite newborn screening early abnormalities in LCI are present in CF patients LCI underscores the need for new treatment approaches to address early lung disease.

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**P4606**

Assessment of ventilation inhomogeneity in patients with alpha-1-antitrypsin deficiency – A useful tool for monitoring early lung disease

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The risk of developing alpha-1-antitrypsin deficiency (α1-AT deficiency) related fatal emphysema during adulthood is high in patients with a PiZZ genotype. Currently, α1-AT deficiency related early lung disease. However it is likely that the early manifestation starts in peripheral room.
airways that are not accessible by spirometry. Multiple Breath Washout (MBW) is a sensitive test for detecting ventilation inhomogeneity (VI) within the peripheral airways that can be quantified by calculating several indices such as the Lung Clearance Index (LCI). We performed a preliminary analysis of an ongoing multi-center study assessing the clinical and prognostic value of measuring the LCI in patients with s-AI deficiency.

Results: 19 controls (7-51 years) and 20 unselected patients with s-AI deficiency (12-72 years) performed 2-3 single MBW N2 using the EasyOne Pro LAB™ (ndd Switzerland) with 100% oxygen and subsequent spirometry. Mean (SD) LCI was 6.5 (0.64) in controls and 9.1 (0.98) in patients with a mean difference (95% CI, p-value) of -2.57 (-3.54-1.60, <0.001) between the groups. Within-test repeatability (CV%) was 5.3% in controls and 6.0% in patients. LCI correlated significantly with age in patients. Assessment of LCI derived from MBW N2 was feasible, reproducible and well tolerated in both, patients and controls. LCI differed significantly between patients and controls. In patients V'Opeak was 80% predicted. From these preliminary data that the LCI reflects presence of VI in patients with s-AI deficiency and may thus be useful for monitoring s-AI deficiency related lung disease.

P4607 Lung clearance index and exercise capacity among children with bronchiectasis
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Background: In paediatric bronchiectasis, there has been limited experience on the relationship between disease severity, as assessed by exercise limitation and lung clearance index (LCI).

Aim: To compare LCI and exercise capacity among children with bronchiectasis.

Method: Fifteen stable children and adolescents with CF, 14 stable children with non-CF bronchiectasis and 15 healthy children and adolescents, participated in maximal incremental cardiopulmonary exercise testing using a cycle ergometer and they had an LCI assessment.

Results: The CF children’s mean age was 13.7 years, mean FEV1 74.4% predicted. The 14 non-CF bronchiectasis children’s mean age was 13.8 years, mean FEV1 75.1% predicted and the healthy children’s mean age was 13.6 years, mean FEV1 94.7%. Among CF patients there was evidence of exercise limitation, with mean Peak Acrobic Capacity (V'Opeak) 62.2% predicted. Among the non-CF bronchiectasis patients there was evidence of exercise limitation, with mean V'Opeak 77.3% predicted. LCI was abnormal in 12/15 (80%) of children. 11/15 patients (73%) had a LCI change in FEV1 (<0.01(3.3)%). Post exercise LCI +1.4(5.5)% in healthy volunteers compared to CSF (-0.2(0.3)%). In those asthma with >20% fall FEV1 (n=3), post salbutamol LCI remained high, 8.0(0.3). Post exercise change in LCI higher in asthma who reported exercise intolerance; No symptoms 2.8(8.5)%; controlled symptoms 5.0(9.3)%; limited 11.3(17.3)%; severe 31.8(18.3)%.

Conclusions: Exercise testing and Multiple Breath Washout measurements can discriminate bronchiectasis from healthy children. However, the burden of the disease is more prominent in children with CF compared with the non-CF bronchiectasis.

P4608 Sensitivity of lung clearance index and chest computed tomography in early lung disease among children with non-CF bronchiectasis
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Background: Lung disease starts before clinical symptoms become prominent, among CF children. Computed chest tomography (CT) is the reference method for identifying structural changes. It has been suggested that the Lung Clearance Index (LCI) is a sensitive marker allowing non-invasive monitoring of lung disease.

Aim: The aim of this prospective study was to investigate the diagnostic accuracy of the LCI in comparison to CT among children with non-CF bronchiectasis, with early lung disease and normal FEV1 (>80% predicted).

Method: MBW and low-dose HRCT were performed in 14 patients (6-21 years) with non-CF bronchiectasis and normal FEV1 (>80% predicted) HRCT scans, LCI and FEV1 were recorded. A modified Bhalla score was used to assess HRCT scans. LCI was assessed with Multiple Breath Washout measurements.

Results: LCI was abnormal in 12/15 (80%) of children. 11/15 patients (73%) demonstrated both, increased LCI and structural changes on CT. 1/15 (6%) had normal results in both measurements. There was a significant linear correlation between CT-score and LCI in 12/15 (87%) of patients; whereas no such correlation was observed between CT-score and FEV1. Sensitivity of the LCI to detect structural lung damage was 85%, whereas specificity of LCI was 50%.

Conclusions: Diagnostic accuracy of the LCI for detecting CF lung disease in patients with normal FEV1, was good when compared to CT. Results indicate that structural changes are unlikely if a normal LCI is measured. The LCI may be a suitable surrogate marker for monitoring progression of lung disease among children with non-CF bronchiectasis.

P4609 Lung clearance index in paediatric exercise induced asthma
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Introduction: Exercise induced bronchoconstriction is reported commonly by asthmatic children. We assessed whether lung clearance index (LCI) can detect it.

Methods: Following symptom questionnaire, spirometry, and Multiple Breath Washout (MBW, Innocor 0.2% SF6), children performed a standard treadmill exercise challenge. One minute post exercise we measured MBW and spirometry. Measures repeated 10 mins post bronchodilatation. If baseline FEV1 decreased >20% baseline, testing only post salbutamol.

Results: 21 asthma patients and 21 healthy controls (13M:8F, mean 12 yrs in both groups). In asthma LCI 7.2(0.7) (mean (SD)) significantly > control 6.8 (0.4), p=0.02. In asthma, post exercise LCI increase by 4.8(10.2)% (Fig 1), with no change in FEV1 (<0.01(3.3)). Post exercise LCI ±4.5% influenced in healthy volunteers (ns). In those asthma with >20% fall FEV1 (n=3), post salbutamol LCI remained high, 8.0(0.3). Post exercise change in LCI higher in asthma who reported exercise intolerance; No symptoms 2.8(8.5)%; controlled symptoms 5.0(9.3)%; limited despite salbutamol 17.8(10.7)% No trend for FEV1; 0.3(9.3)% vs -10.3(1.3)% vs -20.0(3.3)%.

Conclusions: LCI tends to rise following exercise. However rise in LCI was non-significant, consistent with the discrepancy between symptoms and altered lung function in asthmatic children. Change in LCI is greatest in the symptomatic, with largest LCI change in those reporting poor treatment response (not demonstrated by FEV1).

P4610 Hand-held tidal breathing nasal nitric oxide measurement as a targeted case-finding tool for primary ciliary dyskinesia
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Background: Nasal Nitric Oxide (nNO) measurement is a sensitive supplementary tool in diagnosis of Primary Ciliary Dyskinesia (PCD). Tidal Breathing (TB) nNO requires minimal cooperation, has potential as more widespread targeted case-finding tool for PCD in all age groups, and discriminative capacity between PCD and non-PCD has been previously established using stationary nNO analyzer (Martin JK and Nielsen KG. Eur Respir J 2011; 37: 559-565).

Aim: Assess validity of hand-held TBnNO in a selected population.

Methods: TBnNO was measured in PCDs, cystic fibrosis (CF) patients and healthy subjects (HS) using both an electrochemical hand-held device, NIIXO MINO® Nasal equipped with a nasal research application, and two chemiluminiscence stationary nNO analyzer (Marthin JK and Nielsen KG. Eur Respir J 2011; 37: 559-565).

Results: TBnNO values were compared in 41 subjects between 0.3 and 57 years: 15 PCDs, 13 CF patients, and 13 HS. MINO discriminated significantly between PCD and HS (P=0.001) and between CF and PCD (P<0.001).

Conclusions: LCI tends to rise following exercise. However rise in LCI was nonsignificant, consistent with the discrepancy between symptoms and altered lung function in asthmatic children. Change in LCI is greatest in the symptomatic, with largest LCI change in those reporting poor treatment response (not demonstrated by FEV1).
**Conclusion:** Hand-held TlnNO separated significantly between PCD and HS, and between PCD and CF, with cut off value and sensitivity/specificity comparable to those of stationary systems.

**P4611**
Upper and lower airway nitric oxide levels in primary ciliary dyskinesia
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**Introduction:** Patients with primary ciliary dyskinesia (PCD) have low nasal nitric oxide (nNO) and fractional exhaled NO (FeNO). The reasons are unclear but might be related to ciliary function. Analysis of nitric oxide (NO) from different regions of the airway and comparisons with other disease states may guide our understanding.

**Aim:** To compare differential bronchiolar (JNO) and alveolar (CalvNO) NO in patients with PCD, cystic fibrosis (CF), asthma and healthy subjects.

**Methods:** Exhaled NO at different flow rates (50, 100, 200 and 250 mls) and nNO were measured (NIOX flex®, Aerocrine, Sweden) in patients with PCD (n=12), asthma (n=18), CF (n=12) and healthy controls (n=17). JNO and CalvNO were derived using a model of pulmonary NO exchange-dynamics. Results: FeNO50 and nNO were significantly lower in PCD than in healthy subjects, as was JNO2, 271 ppb (228) vs. 965 ppb (963) (p=0.004) (mean SD). However CalvNO was similar between the two, 1.6 ppb (0.5) vs. 2.4 ppb (1.4) (p=0.174). (Table 1 for CF and asthma data)

**Conclusion:** PCD patients have significantly lower JNO but similar CalvNO to healthy controls. As there are no cilia in the alveolar region this might support the hypothesis that NO biosynthesis is coupled to ciliary function. Data collection continues.

**P4612**
Determinants of functional deficits assessed by spirometry and plethysmography in children with bronchial asthma
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Follow-up in asthmatic children is usually performed by spirometry, and hence without consideration of pulmonary hyperinflation. 'Effective' specific follow-up measures resistive changes throughout the whole breath cycle, consequently to changes of resting end-expiratory level.[1]

**Objectives:** Defining the pattern of functional deficits and their changes over time in children with bronchial asthma.

**Methods:** Spirographic measurements performed in 216 asthmatic children (age: 4.0 to 17.9 y) were analyzed retrospectively representing at least 3 annual tests, and providing functional residual capacity (FRCtot), sRaw, volume-time and flow-volume indices (FEV1, FEF25–75, MMEF25–75). Data were expressed as SD-score computed by z-transformation using reference equations. Results: Within the 1207 lung function tests spirometry failed to detect abnormal lung function in 24.1% of tests (plethysmography 1.7%). Bronchial obstruction (> 2SDS) was depicted by sRaw in 93.8%, sRes in 74.6%, MMEF25–75 in 73.8%, MMEF25–75 in 62.1% and FEV1 in 23.7% of tests. Moreover, pulmonary hyperinflation (FRCtot > 2 SDS) was present in 26.7%, mostly combined with obstruction (23.4%). Independent from age at entry pulmonary hyperinflation remained to a certain degree despite treatment (LABA and ICS).

**Conclusions:** Apart from spirometry, follow-up of asthmatic children should include plethysmographic measurements, because changes in static lung volumes influence airway dynamics, mimicking normal flow-volume curves. Moreover, patients with pulmonary hyperinflation are less responsive to standard treatment. [1] Mathys H, Orth U. Respiration 1975: 32:2, 121-134.

**P4613**
Breath by breath specific airway resistance during sleeping in children
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**Rationale:** Panting is the optimal way to minimize the phase lag between the plethysmographic volume compression signal (Vplet) and airflow due to the thermal artifact when measuring specific airway resistance (sRaw). A drawback in children is that the end expiratory lung volume may be observed to increase during the measurement. It is not known to what extent sRaw is altered by the breathing strategy.

**Hypothesis:** The hypothesis was tested here that increased end expiratory lung volume during panting does not impact significantly on sRaw.

**Objective:** To test whether increased end expiratory lung level within the sRaw acquisition results in a systematic trend with time.

**Methods:** sRaw was measured in 10 children panting in a custom made pressure plethysmograph. Thirty measurements (1-5 per subject) that displayed a steady increase in end expiratory lung level throughout at least 4 breaths were reanalyzed breath by breath.

**Results:** Panting frequency (mean ± SD) was 3 ± 1.0.5 Hz. sRaw was found to increase significantly throughout the acquisition (p = 0.006).

**Conclusion:** The progressive increase in end-expiratory lung volume during panting in children is associated with parallel increase in sRaw. This may be explained by non lineairities in the Vplet - airflow relationship while the end expiratory lung volume increases in relation with between inspiratory and expiratory efforts. The clinical relevance is not clear and deserves further evaluation.

**P4614**
Lung function and gender at twelve to thirteen years of age in children born very prematurely
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Lung function abnormalities are common in school aged children born extremely prematurely. Follow up of infants born at 23-28 weeks gestational age from the United Kingdom Oscillation Study (UKOS) showed males had a higher incidence of respiratory morbidity during infancy.

**Aim:** To test the hypothesis that lung function at 12-13 years of age in children born very prematurely would be worse in males than females.

**Methods:** Lung function was assessed in 65 children, the first to date assessed in follow-up of UKOS, which randomised babies to high frequency oscillation or conventional ventilation. Forced expiratory volume in one second (FEV1), forced vital capacity (FVC), FEV1/FVC, residual volume (RV), transfer factor for carbon monoxide (TLCO), functional residual capacity (FRC) and response to cold air challenge (CACH) were assessed. Results were abnormal if two standard deviations (SD) below expected, except RV results which were abnormal if >2 SDs.

**Results:** 29 males and 36 females had been assessed. A greater proportion of males compared to females were oxygen dependant at 28 days (89% versus 69%, p=0.063), had reduced FEV1 (22% versus 3.5%, p=0.036), higher RV (34% versus 5%, p=0.009.17%, but there were no significant differences regarding the proportions with reduced FEV1-FVC (44% versus 31%, p=0.307 (FET p=0.31)) or reduced TLCO (29% versus 50%, p=0.18 (FET p=0.23) or respondence to a CACH (24% versus 26%, p=0.94).

**Conclusion:** Preliminary results suggest that amongst 12-13 year old children born at 23-28 weeks gestation, males have greater airways obstruction than females, but this is not explained by greater airway hyper-reactivity.

**P4615**
Lung function in infants and toddlers after the repair of congenital diaphragmatic hernia (CDH)
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There are only 4 studies on infant pulmonary function testing (IPFT) performed after repair of CDH. We used a wide spectrum of IPFT methods to test babies in whom CDH repair was performed.

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**Table 1. Nitric oxide readings by respiratory disease**

<table>
<thead>
<tr>
<th>Reading</th>
<th>PCD</th>
<th>Healthy</th>
<th>Asthma</th>
<th>CF</th>
</tr>
</thead>
<tbody>
<tr>
<td>nNO (ppb)*</td>
<td>23 (6-61)</td>
<td>856 (536-988) (p&lt;0.001)</td>
<td>769 (560-1126) (p&lt;0.001)</td>
<td>521 (457-697) (p&lt;0.028)</td>
</tr>
<tr>
<td>FeNO50 (ppb)**</td>
<td>9.2 (7.9)</td>
<td>21 (21) (p&lt;0.019)</td>
<td>43 (41) (p&lt;0.001)</td>
<td>15 (11) (p&lt;0.161)</td>
</tr>
<tr>
<td>JNO2 (ppb)**</td>
<td>271 (263)</td>
<td>965 (963) (p&lt;0.004)</td>
<td>764 (542-907) (p&lt;0.03)</td>
<td>5.4 (3.5) (p&lt;0.001)</td>
</tr>
<tr>
<td>CalvNO (ppb)**</td>
<td>1.6 (0.5)</td>
<td>2.4 (1.4) (p&lt;0.174)</td>
<td>2.3 (1.1) (p&lt;0.195)</td>
<td></td>
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</table>

*Median (IQR), **Mean (SD), p values compared to PCD.
We tested 30 infants and toddlers after CDH repair (BW 3.10±0.54 kg [mean±SD]; body length 49.5±2.2 cm). In 4/30 Goretx patch was used (subgroup GORE). Age at testing was 1.32±0.54 (median 1.07) yrs; body weight 9.76±1.25kg (z-score -0.777), body length 78.8±6.7cm (z-score -0.024). The whole-body plethysmography (to measure FRCp and sReff), tidal breathing analysis (tPTEF%tE), baby resistance/compliance (specific Crs) and RTC method (V’maxFRC) were performed. Standard protocols1,2 and proper reference values3,4 were used. FRCp equals 126.5±36.9% pred (P<0.002), sReff reached 109.9±58.9% pred (ns). A parameter of tPTEF%tE decreased (22.2±8.5%). Specific compliance of the respiratory system, rs (Crs/kg) was 14.1±2.3 ml/kPa/kg (76.1±20.1% pred) A value of V’maxFRC reached only 112±4ml/sec (z-score -2.387). Increased value of FRCp was found in GORE subgroup (165.7±51.9 vs. 120.4±31.2, p<0.02).

Our cohort had normal body length but mildly lowered body weight. Neither central airway (aw) obstruction nor restrictive pattern was found. Mild peripheral aw obstruction, mild (secondary) hyperinflation and mildly decreased specific Crs was found.

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References: