improved by >1 kg/m^2, 4 (9%) deteriorated by >1 kg/m^2. Psychological distress was low with 7 (15.6%) having anxiety and 3 (6.7%) depression; 84.4% used 'optimistic acceptance' as their main way of coping, 8.8% used 'avoidance' 2.2% 'distraction', and 2.2% 'hopefulness'.

Conclusion Young people with CF still face daunting problems but are functioning well. There is a need for close monitoring during transition to provide treatment and support to those showing clinical deterioration.

REFERENCE

1 Abott J. *Disability Rehabilitation* 2001;**23**:315.

P183 BURDEN OF ILLNESS IN SCHOOL-AGED PATIENTS WITH CYSTIC FIBROSIS (CF) IN THE UNITED STATES

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Background and objectives The CF disease burden among school-aged children is not well understood. We sought to describe this burden in patients with CF aged 6–17 years in the United States by comparing their healthcare resource utilisation (HCRU) to that of demographically similar controls without CF.

Methods This retrospective study used administrative claims from the Truven MarketScan Medicaid Multi-State (CAID) and Commercial (COMM) databases. Patients with CF aged 6–17 years were identified as having ≥1 inpatient (IP) or ≥2 outpatient (OP) medical claims ≥30 days apart with primary diagnosis of CF (ICD-9-CM: 277.0x) between 2010 and 2014. Other inclusion criteria were ≥12 months of continuous medical and pharmacy coverage and ≥1 CF-related healthcare encounter during the most recent year of data. Patients were matched 1:3 to non-CF controls by age, gender, race (CAID cohort), geographic region (COMM cohort), insurance plan type and enrollment (calendar year). IP admissions, OP visits and medication use from the most recent year of data (2010–2014) were compared between patients and controls, overall and by age group (6–11 and 12–17 years), using bivariate statistics; chi-square tests were used for categorical variables and *t* tests and ANOVA for continuous variables.

Results The CAID cohort included 1264 patients with CF and the COMM cohort 2400; all were matched 1:3 to controls (mean [SD] age of patients and controls: CAID, 11.4 [3.5]; COMM, 11.9 [3.5]). Annual hospitalisation rates were 22-fold (CAID) to 32-fold (COMM) higher in the CF cohorts, with lengths of stay nearly twice that of matched controls (**Table**). Annual OP visit rates were 3.1-fold (CAID) and 3.5-fold (COMM) higher in the CF cohorts, and patients filled 5 times as many unique medications and 10 times as many total prescriptions per year as controls. While patients with CF aged 12–17 years generally had higher HCRU than those aged 6–11, trends and magnitude difference vs controls within each age group were similar.

Conclusion HCRU was higher in patients with CF aged 6–17 years than in demographically similar children without CF, illustrating significant disease burden and a need for better treatment options for this population.

Sponsored by Vertex Pharmaceuticals Incorporated

Please refer to page A272 for declarations of interest in relation to abstract P183.

P184 CALCULATION OF CONDUCTIVE INHOMOGENEITY IN CHILDREN WITH SEVERE CF LUNG DISEASE: WHICH METHOD WORKS?

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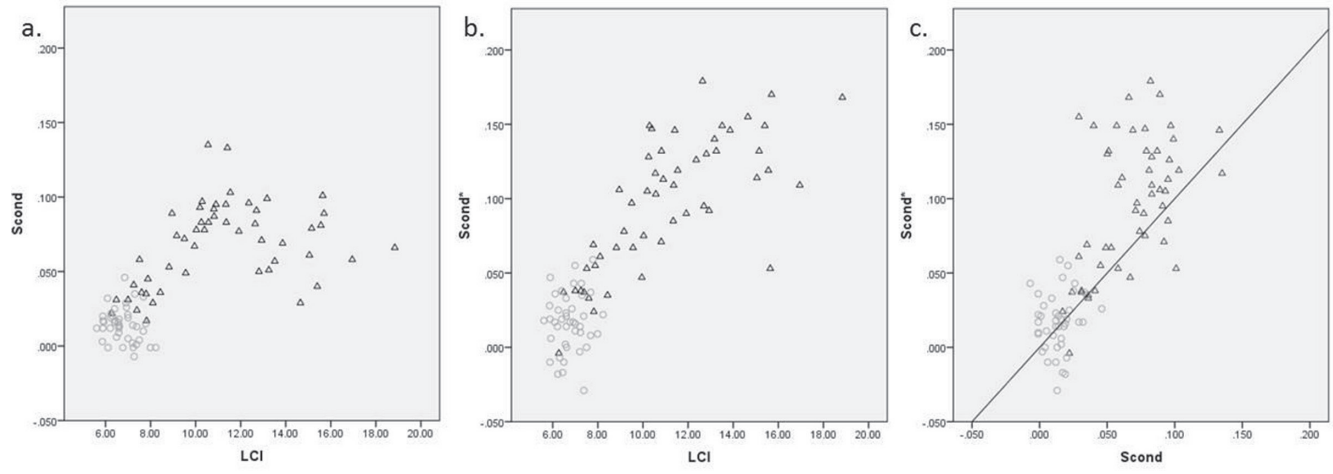
Introduction Convection Dependent Inhomogeneity (CDI, a measure of ventilation inequality among larger lung units) quantified by Scond cannot be assessed in subjects with severe

Abstract P183 Table 1

	Total Cases	Total Controls	Age 6–11 y Case	Age 6–11 y Control	Age 12–17 y Case	Age 12–17 y Control
Medicaid (CAID), n	1264	3792	642	1926	622	1866
Proportion with at least 1 IP admission, n (%)	521 (41.2)	107 (2.8)	202 (31.5)	32 (1.7)	319 (51.3)	75 (4.0)
Annual IP admissions, mean (SD)	0.87 (1.49)	0.04 (0.25)	0.55 (1.10) [a]	0.02 (0.17) [a]	1.20 (1.76)	0.06 (0.30)
LOS per admission, mean (SD), days	10.1 (9.0)	6.8 (8.3)	9.5 (6.2) [b]	6.4 (7.7) [b]	10.5 (10.3) [a]	6.9 (8.6) [a]
Annual OP office visits, mean (SD)	9.9 (8.0)	3.2 (3.9)	9.6 (7.6)	3.3 (4.1)	10.2 (8.4)	3.0 (3.7)
Annual total prescriptions filled, mean (SD)	67.3 (52.6)	7.2 (13.4)	65.5 (54.0)	6.6 (11.3)	69.1 (51.1)	7.8 (15.1)
Annual unique prescriptions filled, mean (SD)	15.8 (8.5)	3.2 (4.1)	14.5 (7.3)	3.0 (3.8)	17.1 (9.4)	3.4 (4.3)
Commercial (COMM), n	2400	7200	1075	3225	1325	3975
Proportion with at least 1 IP admission, n (%)	816 (34.0)	107 (1.5)	270 (25.1)	32 (1.0)	546 (41.2)	75 (1.9)
Annual IP admissions, mean (SD)	0.64 (1.21)	0.02 (0.20)	0.40 (0.88) [b]	0.01 (0.10) [b]	0.85 (1.39)	0.03 (0.26)
LOS per admission, mean (SD), days	8.4 (6.3)	4.5 (5.9)	7.5 (4.8)	3.6 (4.2)	8.8 (6.9)	4.9 (6.5)
Annual OP office visits, mean (SD)	9.9 (6.6)	2.8 (3.5)	9.4 (6.0)	2.7 (2.9)	10.3 (7.0)	2.8 (3.8)
Annual total prescriptions filled, mean (SD)	39.8 (31.4)	3.6 (7.0)	37.5 (29.0)	2.9 (5.6)	41.7 (33.1)	4.1 (7.9)
Annual unique prescriptions filled, mean (SD)	11.6 (7.0)	2.0 (2.8)	10.5 (6.1)	1.7 (2.3)	12.5 (7.5)	2.2 (3.0)

P value <0.001 for all comparisons (case vs control), unless otherwise noted. [a]*P* <0.01; [b]*P* <0.02. LOS =length of stay.



Abstract P184 Figure 1 CF triangles, Control circles. A. Scond vs LCI b, Scond* vs LCI c. Scond* vs Scond with line of equivalence

ventilation inhomogeneity (VI) as assumptions underlying the calculation are invalid; an alternate index that has been suggested is Scond.[1]

Aim To compare these two methods of CDI assessment in CF children

Methods Children with cystic fibrosis (CF; 67) and healthy controls (61) performed multiple breath washout with sulphur hexafluoride measured using mass spectrometry. Scond was calculated from 1.5 to 6 turnovers and Scond* from breath 2 to 3 turnovers.

Results All measures of VI were significantly higher for CF vs control, mean difference: LCI 4.0, Scond 0.054, Scond* 0.081.

In CF, LCI correlated better with Scond* than Scond (See figure: correlation coefficient LCI vs. Scond* 0.75; LCI vs. Scond 0.42). If children with moderate-severe VI (LCI > 11) were excluded there was an improved correlation for both relationships (correlation coefficient LCI vs. Scond 0.83; LCI vs. Scond* 0.86).

An asymptote for the Scond vs LCI relationship was at Scond 0.07 and Scond* 0.13.

Conclusion Scond* quantifies the mechanism of VI in moderate to severe lung disease, but it may reach asymptote in very severe VI.

REFERENCE

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P185 SLEEP DISORDERED BREATHING IN CHILDREN WITH SPINA BIFIDA. TIME TO SCREEN?

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Background Spina bifida is associated with sleep disordered breathing (SDB) particularly when associated with Arnold-Chiari malformations. Studies suggest that moderate/severe sleep apnoea is present in up to a third of spina bifida patients (Patel 2015) and yet there are no national guidelines that recommend screening for SBD in children with spina bifida. There is evidence to suggest that many children present late and this can be associated with unnecessary morbidity and even mortality (Kirk 1999).

Aim To assess the prevalence of SDB in children with spina bifida, presenting through clinical presentation alone in the West of Scotland and to explore whether there is a case for screening all children with spina bifida for SDB.

Method The database of the Spina Bifida Association Scotland and clinical records from the regional centre in the Royal Hospital for Children, Glasgow were used to identify all children with spina bifida in the West of Scotland. The level of the spinal lesion, presence of an Arnold-Chiari malformation or ventriculoperitoneal shunt was established, as was the number who had had sleep studies performed and who had required ventilator support.

Results 108 children were identified; 44/108 (40%) had an Arnold-Chiari malformation (1 type I, 43 type II); 64/108 had lumbar abnormalities, 14/108 lumbosacral, 14/108 thoracolumbar, 9/108 sacral and 4/108 thoracic. 52/108 had a VP shunt at some point. Only 14 children had presented with clinical symptoms that lead to a sleep study being undertaken (snoring 7, apnoeas 7, cough/wheeze 2, restlessness at night 2, morning headache 2). 5 children had mixed central and obstructive apnoeas, 1 obstructive sleep apnoea, 2 hypoventilation. 8 children went on to require non-invasive mask ventilation of these 7/8 had an Arnold-Chiari malformation ($p = 0.005$), 7/8 had a previous VP shunt ($p = 0.02$), 5/8 had lumbar abnormalities and 3/8 thoraco-lumbar.

Conclusion Clinical presentation alone only identifies a small proportion of cases of SDB in children with spina bifida, with a high proportion of these requiring intervention. We remain concerned that there are many children with spina bifida with undiagnosed SDB who may benefit from treatment, particularly those with Arnold-Chiari malformations and therefore that screening is indicated.

P186 INCIDENCE AND OUTCOME OF CONGENITAL LUNG AGENESIS IN THE NORTH OF ENGLAND

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