



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

ventilation inhomogeneity (VI) as assumptions underlying the calculation are invalid; an alternate index that has been suggested is Scnd.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. Scnd was calculated from 1.5 to 6 turnovers and Scnd* from breath 2 to 3 turnovers.

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

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

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

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

REFERENCE

¹ Verbanck. *Respiratory Physiology & Neurobiology*, 2013.

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 SDB 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 ventriculo-peritoneal 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/whoeze 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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