

SUPPLEMENTARY METHODS

Patient selection -

University Hospital Southampton NHS Foundation Trust is one of three nationally commissioned PCD diagnostic centres in the UK[1]. We undertake PCD diagnostics on patients of all ages primarily from across the South and South-West of the UK.

Of patients referred between June 2006 and January 2014, 282 were aged 6 years or over and were able to perform nasal nitric oxide testing. Children under 6 years were excluded as there is evidence that nNO levels are lower in healthy children in this age group and, therefore, diagnostic test summary statistics might be different for this population.

Nasal NO testing -

Testing was performed on the NIOX Flex stationary chemiluminescent nitric oxide analyser at a sampling rate of 5ml/s as per manufacturer instructions. Sampling was undertaken during velum closure by either breath hold or oral exhalation against resistance. Visual representation of sampling trace was observed and reading taken at plateau. Highest value was recorded from 3 satisfactory tests. Level in parts per billion (ppb) was converted to nanolitres per minute (nl/min) to allow comparison with other analysers ($nl/min = level\ in\ ppb \times sampling\ rate\ in\ l/min$).

Diagnosis of PCD -

Patients with a supportive clinical history were considered to be PCD positive if ciliary beat frequency or pattern was abnormal alongside abnormal ciliary ultrastructure on electron microscopy [2]. Patients were also considered PCD positive if ultrastructure was normal but characteristic ciliary beat pattern abnormalities were consistently seen on two healthy samples taken at different times. Any patients where the diagnosis was "indeterminate" were excluded from this analysis; this included patients where repeated nasal brushings were insufficient for analysis or diagnostic testing was not conclusive.

References

1. Lucas JS, Burgess A, Mitchison HM *et al*. Diagnosis and management of primary ciliary dyskinesia. Arch Dis Child. 2014 Apr 25;
2. Jackson CL, Behan L, Collins SA, et al. Accuracy of diagnostic testing in primary ciliary dyskinesia. Eur Respir J 2015;*In Press* doi: 10.1183/13993003.00749-2015.

	nNO above 77nl/min	nNO below 77nl/min	Total
PCD positive	2	29	31
PCD negative	211	40	251

Table S1. Numbers of PCD positive and negative patients with nasal nitric oxide (nNO) above or below screening cut-off of 77 nanolitres per minute (nl/min)