Dear Editor,

We thank Professor Greenough and Dr Lunt for their interest1 in our manuscript,2 and for the suggestion that changes in pulmonary vascular volumes may lead to airflow obstruction. In this group of sickle cell diseased (SCD) children and controls, we used carbon monoxide transfer (DLCO) related to pulmonary blood flow (Qpeff) at rest and on exercise as a surrogate for pulmonary capillary blood volume.3 DLCO corrected for haemoglobin and surface area was significantly higher in SCD at rest, but only by about 10%, while Qpeff was 15–20% higher. DLCO:Qpeff, therefore, was significantly lower in SCD at rest and remained so at all exercise stages, thus implying lack of normal recruitment and distension of the pulmonary microcirculation, and suggesting that at rest at least, the microvasculature is unlikely to be contributing to airflow obstruction, a result which contrasts with their findings.4 This discrepancy is unexplained. In the meantime, we agree that further mechanistic research is needed to try to understand why airflow obstruction develops and how it contributes to the pathophysiology of lung disease in these children.

Suzanne Crowley,1 Rifat A Chaudry,1,2
Mark Rosenthal,1 Andrew Bush2,3
1Department of Paediatrics, St George’s Hospital, London, UK
2Department of Paediatric Respiratory Medicine, Royal Brompton Hospital, London, UK
3Department of Paediatrics, Imperial College, London, UK
Correspondence to Dr Suzanne Crowley, Section for Paediatric Lung and Allergic Diseases, Oslo Universitetssykehus, Rikshospitalet, Oslo 0027, Norway; suzzcro@ous-hf.no
Contributors SC and AB drafted the response, and all approved the final draft.
Competing interests None.
Provenance and peer review Not commissioned; internally peer reviewed.

REFERENCES