MedicalXpress

For children with sickle cell disease, lung disease is part of the package

May 18 2008

Children with sickle cell disease (SCD) have a significantly sharper decline in lung function with age when compared to other children of the same race and age. Furthermore, that loss of function appears to be linked to a restrictive rather than obstructive pattern, contrary to previous research that has focused on obstructive or asthma-like patterns in loss of lung function with sickle cell disease. The research will be presented at the American Thoracic Society's 2008 International Conference in Toronto on Sunday, May 18.

"The restrictive pattern of decline is supportive of early injury or inflammation resulting in progressive changes in lung volumes across age," said lead researcher Joanna MacLean, M.D., of the department of respiratory medicine at Children's Hospital at Westmead in Australia. "We expected that children with sickle cell disease would show greater loss of lung function than other children, but this had never been quantified, nor was the pattern of decline clear."

To determine the patterns of loss of lung function, the researchers analyzed 1,357 lung function results that were completed between January 1989 and January 2005 on from 413 children with SCD during routine sickle cell clinical visits. Lung volume measurements were also included for 1,129 records.

They found that over time, there was a significant decline in lung function shown by a decline in the percent predicted values for all spirometry measures except FEV1/FVC ratio, a marker of airway

obstruction. The pattern of decline of lung volume confirmed a restrictive pattern with an average loss of 2 percent per year of total lung capacity.

This is the first study to examine loss of lung function in children with sickle cell disease over time in a large number of children. “Most studies of lung function in children with sickle cell disease are either cross-sectional or have limited longitudinal follow-up,” said Dr. MacLean. “The strength of our study is that data was collected from a large number of children with sickle cell disease across childhood.”

They also found that of children with SCD, hemoglobin-beta genotype SS was associated with an increased risk of loss of lung function across childhood. The hemoglobin SS genotype is more common than hemoglobin SC genotype (one in 600 versus one in 800 life births) and is associated with more complications than hemoglobin SC disease. This pattern of decline in lung function may simply reflect a greater risk of lung injury in this group, but alternatively may also suggest differences in response to lung injury or lung repair with different genotypes of sickle cell disease.

“Our findings confirm that lung disease in SCD begins in childhood,” explained Dr. MacLean. “Using statistical modeling, we are able to predict the rate of decline of lung function. These results can be used as a baseline against which results from intervention studies can be compared.”

While changes in treatment over the last 20 years have led to a significant reduction in overall mortality and morbidity for children with SCD, prospective longitudinal studies focusing on the identification and treatment of sickle cell associated lung disease are needed to continue to improve the long term health of children and adults with sickle disease.

“The results of this project emphasize the need for further investigation into the pathophysiology and treatment of lung disease in children with sickle cell disease,” concluded Dr. MacLean.

Source: American Thoracic Society