

SCIOS: A study of Airway Resistance by Impulse Oscillometry System (IOS) in Children with Sickle Cell Disease (SCD)

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Synopsis:

Hypothesis: Airway resistance measured by IOS is higher in 3-6 year old sickle cell patients compared to controls without Sickle cell disease (SCD).

Specific Aims:

1. To determine if airway resistance is higher in 3-6 year old children with SCD compared to those without SCD.
2. To determine if bronchodilator response is greater in 3-6 year old children with SCD vs. those without SCD

Sickle cell disease (SCD) is one of the most prevalent genetic disorders in the US and affects more than 50,000 Americans. It is associated with significant pulmonary complications including Acute Chest Syndrome (ACS), Pulmonary Hypertension (PHT) and Sickle Cell Chronic Lung Disease (SCLD). Patients with SCD often exhibit pulmonary function abnormalities in the form of obstructive and/or restrictive patterns, as well as airway hyper-reactivity. Recent studies have shown increased association between ACS and Asthma in SCD. Lower airway obstruction and reversibility is more common in younger patients with SCD, while restrictive changes are seen more often in older children and adults with SCD and are thought to result from repeated episodes of ACS. It is not known if pulmonary function abnormalities start early in life or if they predict future clinical outcome.

The classic techniques of measuring lung function- spirometry and plethysmography requires the cooperation of young patients and is often not feasible in pre-school children. On the other hand, IOS is a relatively easy technique, and provides an alternate way to check lung function. Using this technique, airway resistance and response to bronchodilator can be accurately obtained, which can give an estimate of airway obstruction.

The objective of this study is to compare airway resistance and bronchodilator response in 3 to 6 year old SCD patients with age, gender and racially matched normal controls, to determine if airway obstruction is present early in life. To the best of our knowledge, this is the first study to look at pulmonary function by IOS in children between 3 and 6 year old with SCD.