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Early detection of cystic fibrosis lung disease

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Cystic fibrosis lung disease starts very early in life, and is characterised by a cycle of infection and inflammation that causes progressive lung damage and ultimately leads to respiratory failure. The goal of treatment of young children with cystic fibrosis is to identify this lung disease early and intervene at an early stage in this cycle in an attempt to prevent progression. This poses a new challenge both to the clinician looking after these patients and also for the researcher developing new interventions, as the traditional outcome measures for CF lung disease such as Shwachman scores, plain chest radiography, and spirometry were designed to track moderate-to-severe disease but are less helpful in detection of early or mild disease. The importance of this challenge cannot be overstated. For the clinician it is necessary to identify those children who may benefit from more aggressive therapy, and target that therapy to them. For the researcher it is necessary to find outcome measures that reliably detect early disease and which are sensitive and responsive enough so that intervention studies can be designed.

The two most commonly cited alternative measures are high resolution computed tomography (CT) scans, and measures of ventilation distribution (gas mixing) from multiple breath inert gas washout (MBW) tests. Neither test is new, with the early literature for both investigations now stretching back for more than twenty years. Research in both fields is continuing rapidly. In the case of CT scanning developments have focused upon refinement of scoring systems to quantify changes of early structural disease, and on development of new data acquisition algorithms that can reduce the radiation exposure required for the scan. Development of the MBW technique is still at an earlier stage, as there is still no completely accepted commercial equipment available. Development is therefore still focused on standardisation of data collection and interpretation.

It should be emphasised that these two investigations monitor different aspects of CF lung disease and are not analogous. CT scanning is a record of structural lung change that can detect early bronchiectasis, mucus plugging, and indirectly detect small airways disease. The interpretation of MBW is more controversial. There are good theoretical reasons why the MBW test should be a more sensitive marker of peripheral airways disease than most other lung function tests. However, that does not mean that an abnormal MBW result necessarily equates to small airways disease, as there are many other pathologies (for example mucus production in the airways) that can also result in abnormal ventilation distribution. It is safer to state that an abnormal MBW result simply represents inefficient gas mixing rather than speculate upon the exact pathology that leads to this.

These controversies should not necessarily represent major obstacles to the clinical application of these tests. Ultimately, the need for both the clinician and the researcher is for a test that is relatively simple and cheap to perform, and that involves minimal or ideally no risk to the child. In addition, the test should be repeatable, should respond to successful interventions, and should ultimately reflect the progression of the disease. From current data it appears likely that in the future both tests will be used in conjunction to monitor the progress of young children with cystic fibrosis.