

Improvement in clinical markers in CF patients using a reduced glutathione regimen: An uncontrolled, observational study.

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CFTR mutation, which causes cystic fibrosis (CF), has also recently been identified as causing glutathione system dysfunction and systemic deficiency of reduced glutathione (GSH). Such dysfunction and deficiency regarding GSH may contribute to the pathophysiology of CF. We followed 13 patients (age range 1-27 years) with cystic fibrosis who were using a regimen of reduced glutathione (GSH), including oral glutathione and inhaled buffered glutathione in an uncontrolled, observational study. Dosage ranged from 66-148 mg/kg/day in divided doses, and the term examined was the initial 5.5 months of GSH use (45 days of incrementally adjusted dose, plus 4 months of use at full dosage). Baseline and post-measurements of FEV1 percent predicted, BMI percentile, and weight percentile were noted, in addition to bacterial status and pulmonary exacerbations. Significant improvement in the following clinical parameters was observed: average improvement in FEV1 percent predicted (N=10) was 5.8 percentage points ($p<0.0001$), average weight percentile (N=13) increased 8.6 points ($p<0.001$), BMI percentile (N=11) improved on average 1.22 points ($p<0.001$). All patients improved in FEV1 and BMI, if measured in their case; 12 of 13 patients improved in weight percentile. Positive sputum cultures of bacteria in 11 patients declined from 13 to 5 ($p<0.03$) with sputum cultures of *Pseudomonas aeruginosa* becoming negative in 4 of 5 patients previously culturing PA, including two of three patients chronically infected with PA as determined by antibody status. Use of a daily GSH regimen appears to be associated in CF patients with significant improvement in lung function and weight, and a significant decline in bacteria cultured in this uncontrolled study. These findings bear further clinical investigation in larger, randomized, controlled studies.

