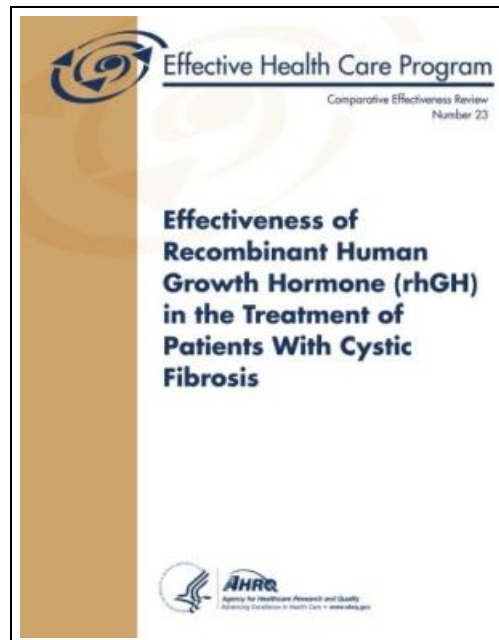


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Createspace, United States, 2013. Paperback. Book Condition: New. 279 x 216 mm. Language: English . Brand New Book ***** Print on Demand *****.Cystic fibrosis (CF) is the second most common life-shortening, childhood-onset genetic disease in the United States, affecting approximately 30,000 people in the Nation. The gene responsible for CF encodes the cystic fibrosis transmembrane regulator (CFTR) protein, which regulates sodium and chloride transport across epithelial membranes. This affects nearly all exocrine glands, with abnormally viscous mucus and excessive secretions. The dominant clinical features are chronic lung disease and pancreatic insufficiency with poor nutrition and growth. Treatment advances in CF over the past 25 years have improved measures of nutrition, pulmonary function, and mortality. Growth and nutritional indexes may be predictive of future pulmonary function in children with CF. It has been suggested that improvement of linear growth in children with CF may allow more lung mass and better pulmonary function, independent of improved weight gain. Both poor weight and shorter height have also been shown to be independently associated with increased morbidity and mortality in CF patients in some studies. Recombinant human growth hormone (rhGH) is an anabolic agent with a wide variety of actions. It has been investigated for the treatment of CF because of the decreased growth measures and increased energy expenditures in CF patients. This Review, prepared by the University of Connecticut/Hartford Hospital Evidence-based Practice Center (EPC), examines the benefits and harms associated with using rhGH in patients with CF. The key questions examined are: KQ 1: In patients with CF, does treatment with rhGH as an adjuvant to usual care improve intermediate outcomes, including pulmonary function; growth (height, weight, lean body mass [LBM], protein turnover), exercise tolerance, and bone mineralization, compared with usual care alone? KQ 2: In patients with CF, does treatment with rhGH...



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