



THE CF SUPPLEMENT

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NUTRITION AND CYSTIC FIBROSIS: PUBLICATIONS FROM 2009



This issue of The CF Supplement is the annual listing of nutrition publications appearing in peer-reviewed journals in the previous year. The papers were identified through a search of the Pub Med database, are sorted by topic, and listed in alphabetical order by the last name of the first author.

FATTY ACIDS

Persistence of essential fatty acid deficiency in cystic fibrosis despite nutritional therapy. Aldámiz-Echevarría L, et al. *Pediatr Res.* 2009 Nov;66(5): 585-9.

Association of cholesterol oxidation and abnormalities in fatty acid metabolism in cystic fibrosis. Iuliano L, et al. *Am J Clin Nutr.* 2009 Sep; 90(3):477-84.

Increased tissue arachidonic acid and reduced linoleic acid in a mouse model of cystic fibrosis are reversed by supplemental glycerophospholipids enriched in docosahexaenoic acid. Mimoun M, et al. *J Nutr.* 2009 Dec;139(12):2358-64.

Prevalence of dyslipidemia in adults with cystic fibrosis. Rhodes B, et al. *J Cyst Fibros.* 2010 Jan; 9(1):24-8.

Lipid metabolism in cystic fibrosis. Worgall TS. *Curr Opin Clin Nutr MetabCare.* 2009 Mar;12(2):105-9. [Review.](#)

FOUNDATION AND SOCIETY PUBLICATIONS

Abstracts of the 23rd Annual North American Cystic Fibrosis Conference, Minneapolis, Minnesota, USA, October 15-17, 2009. *Pediatr Pulmonol Suppl.* 2009;32:109-453.

Abstracts of the 32nd European Cystic Fibrosis Conference. June 10-13, 2009. Brest, France. *J Cyst Fibros.* 2009 Jun;8 Suppl 2:S1-112.

Cystic Fibrosis Foundation evidence-based guidelines for management of infants with cystic fibrosis. Borowitz D, et al. *J Pediatr.* 2009 Dec; 155(6 Suppl):S73-93.

Cystic Fibrosis Foundation practice guidelines for the management of infants with cystic fibrosis transmembrane conductance regulator-related metabolic syndrome during the first two years of life and beyond. Borowitz D, et al *J Pediatr.* 2009 Dec;155(6 Suppl):S106-16.

Improving the care of infants identified through cystic fibrosis newborn screening. Marshall BC, et al. *J Pediatr.* 2009 Dec;155(6 Suppl):S71-2.

Management of infants with cystic fibrosis: a summary of the evidence for the cystic fibrosis foundation working group on care of infants with cystic fibrosis. Robinson KA, et al. *J Pediatr.* 2009 Dec;155(6 Suppl):S94-S105. [Review.](#)

ENDOCRINE: Bone and Diabetes

Is cystic fibrosis-related diabetes an apoptotic consequence of ER stress in pancreatic cells? Ali BR, et al. *Med Hypotheses.* 2009 Jan;72(1):55-7.

A susceptibility gene for type 2 diabetes confers substantial risk for diabetes complicating cystic fibrosis. Blackman SM, et al. *Diabetologia.* 2009 Sep; 52(9):1858-65.

Diabetes as a determinant of mortality in cystic fibrosis. Chamnan P, et al. *Diabetes Care.* 2010 Feb; 33(2):311-6.

Growth in children with cystic fibrosis-related diabetes. Cheung MS, et al. *Pediatr Pulmonol.* 2009 Dec;44(12):1223-5.

Dawn of the "bone phenotype" in cystic fibrosis. Chotirmall SH, et al. *Pediatrics.* 2009 Feb; 123(2):e353; author reply e353-4.

SPECIAL POINTS OF INTEREST:

- *See the section "Foundation or Society Publications" for papers describing the care of newborns who have CF.*
- *Many new papers focusing on either diabetes or bone disease are listed under the heading "Endocrine."*
- *The "Gastrointestinal" section contains papers focused on liver disease, pancreatitis, or FDA approved enzymes.*

Volume 8, Issue 2:

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Relation of bone mineral density with clinical and laboratory parameters in pre-pubertal children with cystic fibrosis. Cobanoglu N, et al. *Pediatr Pulmonol.* 2009 Jul;44(7):706-12.

Bisphosphonates for osteoporosis in people with cystic fibrosis. Conwell LS, et al. *Cochrane Database Syst Rev.* 2009 Oct 7;(4):CD002010. [Review.](#)

Diabetes mellitus and bone disease in cystic fibrosis. Curran DR, et al. *Semin Respir Crit Care Med.* 2009 Oct;30(5):514-30. [Review.](#)

Validation of insulin secretion indices in cystic fibrosis patients. Hammana I, et al. *J Cyst Fibros.* 2009 Dec;8(6):378-81.

Early glucose abnormalities in cystic fibrosis are preceded by poor weight gain. Hameed S, et al. *Diabetes Care.* 2010 Feb;33(2):221-6.

Use of the insulin pump in treat cystic fibrosis related diabetes. Hardin DS, et al. *J Cyst Fibros.* 2009 May; 8(3):174-8.

Cystic fibrosis transmembraneconductance regulator (CFTR) regulates the production of osteoprotegerin (OPG) and prostaglandin (PG) E2 in human bone. Le Heron L, et al. *J Cyst Fibros.* 2010 Jan;9(1):69-72.

Cystic fibrosis related diabetes mellitus - Diagnostic and management challenges. Lek N, et al. *Curr Diabetes Rev.* 2009 Dec 23. [Epub ahead of print]

Well-nourished cystic fibrosis patients have normal mineral density, but reduced cortical thickness at the forearm. Louis O, et al. *Osteoporos Int.* 2009 Feb; 20(2):309-14.

Bone and body composition analyzed by dual-energy x-ray absorptiometry (DXA) in clinical and nutritional evaluation of young patients with Cystic Fibrosis: a cross-sectional study. Lucidi V, et al. *BMC Pediatr.* 2009 Sep 28;9:61.

Diagnosis of diabetes. What about cystic fibrosis? Marchetti F, et al. *BMJ.* 2009 Dec 31;339:b5644. doi: 10.1136/bmj.b5644.

Mechanisms of glucose intolerance in cystic fibrosis. Mohan K, et al. *Diabet Med.* 2009 Jun;26(6):582-8.

Abnormal glucose tolerance in CF-when should we offer diabetes treatment? Moran A. *Pediatr Diabetes.* 2009 May;10(3):159-61.

Cystic fibrosis-related diabetes: current trends in prevalence, incidence, and mortality. Moran A, et al. *Diabetes Care.* 2009 Sep;32(9):1626-31.

Cystic fibrosis-related diabetes therapy study group. Insulin therapy to improve BMI in cystic fibrosis-related diabetes without fasting hyperglycemia:results of the cystic fibrosis related diabetes therapy trial. Moran A, et al. *Diabetes Care.* 2009 Oct;32(10):1783-8.

Cystic fibrosis-related diabetes: from CFTR dysfunction to oxidative stress. Ntimbane T, et al. *Clin Biochem Rev.* 2009 Nov;30(4):153-77.

Severe bone demineralisation is associated with higher mortality in children with cystic fibrosis. O'Reilly R, et al. *Ir Med J.* 2009 Feb;102(2):47-9.

Validation of continuous glucose monitoring in children and adolescents with cystic fibrosis: a prospective cohort study. O'Riordan SM, et al. *Diabetes Care.* 2009 Jun;32(6):1020-2.

Management of cystic fibrosis-related diabetes in children and adolescents. O'Riordan SM, et al. *Pediatr Diabetes.* 2009 Sep;10 Suppl 12:43-50. [Review.](#)

The prevalence of osteoporosis, osteopenia, and fractures among adults with cystic fibrosis: a systematic literature review with meta-analysis. Paccou J, et al. *Calcif Tissue Int.* 2010 Jan;86(1):1-7.

Routine screening for cystic fibrosis-related diabetes. Peckham D. *J R Soc Med.* 2009 Jul;102 Suppl 1:36-9. [Review.](#)

The efficacy of calcitriol therapy in the management of bone loss and fractures: a qualitative review. Peppone LJ, et al. *Osteoporos Int.* 2009 Dec 4. [Epub ahead of print]

Bone mineral and body composition alterations in paediatric cystic fibrosis patients. Reix P, et al. *Pediatr Radiol.* 2010 Mar;40(3):301-8.

Diagnosis and management of cystic fibrosis-related diabetes in adults. Richards G. *Nurs Stand.* 2009 Mar 25-31;23(29):35-9.

Update on cystic fibrosis-related bone disease: a special focus on children. Sermet-Gaudelus I, et al. *Paediatr Respir Rev.* 2009 Sep;10(3):134-42.

Osteoclast function, bone turnover and inflammatory cytokines during infective exacerbations of cystic fibrosis. Shead EF, et al. *J Cyst Fibros.* 2010 Mar;9 (2):93-8.

'Old' bones in young bodies: the tale of cystic fibrosis. Sparks AA, et al. *Curr Opin Endocrinol Diabetes Obes.* 2009 Dec;16(6):407-14. [Review.](#)

Endocrine parameters of cystic fibrosis: back to basics. Stalvey MS, et al. *J Cell Biochem.* 2009 Oct 1;108(2):353-61. [Review.](#)

Insulin-like growth factor-I levels predict weight, height and protein catabolism in children and adolescents with cystic fibrosis. Switzer M, et al. *Pediatr Endocrinol Metab.* 2009 May;22(5):417-24.

Nutritional decline in cystic fibrosis related diabetes: the effect of intensive nutritional intervention. White H, et al. *J Cyst Fibros.* 2009 May;8(3):179-85.

Demographics of glucose metabolism in cystic fibrosis. van den Berg JM, et al. *J Cyst Fibros.* 2009 Jul; 8(4):276-9.

Cystic fibrosis related diabetes. Zirbes J, et al. *Paediatr Respir Rev.* 2009 Sep;10(3):118-23; quiz 123.

GASTROINTESTINAL

Genetic modifiers of liver disease in cystic fibrosis. Bartlett JR, et al. *JAMA.* 2009 Sep 9; 302(10):1076-83.

Abnormalities and disease in cystic fibrosis: epidemiology and outcomes through adulthood. Bhardwaj S, et al. *Hepatobiliary J Clin Gastroenterol.* 2009 Oct;43(9):858-64.

Co-morbidity of cystic fibrosis and celiac disease in Scandinavian cystic fibrosis patients. Fluge G, et al. *J Cyst Fibros.* 2009 May;8(3):198-202.

Symptomatic pancreatitis in patients with cystic fibrosis. Gooding I, et al. *Am J Gastroenterol.* 2009 Jun;104(6):1519-23.

Wilson disease as a cause of liver injury in cystic fibrosis. Kotalová R, et al. *J Cyst Fibros.* 2009 Jan; 8(1):63-5.

Hepatobiliary disease in patients with cystic fibrosis. Moyer K, et al. *Curr Opin Gastroenterol.* 2009 May; 25(3):272-8. [Review.](#)

Pancreatic enzyme replacement therapy for young cystic fibrosis patients. Munck A, et al. *J Cyst Fibros.* 2009 Jan;8(1):14-8.

Hepatocellular carcinoma complicating cystic fibrosis related liver disease. O'Donnell DH, et al. *J Cyst Fibros.* 2009 Jul;8(4):288-90.

Application of the glucose hydrogen breath test for the detection of bacterial overgrowth in patients with cystic fibrosis—a reliable method? Schneider AR, et al. *Dig Dis Sci.* 2009 Aug;54(8): 1730-5.

Ursodeoxycholic acid treatment in patients with cystic fibrosis at risk for liver disease. Siano M, et al. *Dig Liver Dis.* 2009 Dec 22. [Epub ahead of print]

Efficacy and safety of Creon 24,000 in subjects with exocrine pancreatic insufficiency due to cystic fibrosis. Trapnell BC, Maguiness K, et al. *J Cyst Fibros.* 2009 Dec;8(6):370-7.

Gastric acid inhibition for fat malabsorption or gastroesophageal reflux disease in cystic fibrosis: longitudinal effect on bacterial colonization and pulmonary function. van der Doef HP, et al. *J Pediatr.* 2009 Nov;155(5):629-33.

Exocrine pancreatic function testing in patients with cystic fibrosis and pancreatic sufficiency: a correlation study. Weintraub A, et al. *J Pediatr Gastroenterol Nutr.* 2009 Mar;48(3):306-10.

EUR-1008 pancreatic enzyme replacement is safe and effective in patients with cystic fibrosis and pancreatic insufficiency. Wooldridge JL, et al. *J Cyst Fibros.* 2009 Dec;8(6):405-17.

GROWTH AND ENERGY

Growth and long-term lung function in cystic fibrosis: a longitudinal study of patients diagnosed by neonatal screening. Assael BM, et al. *Pediatr Pulmonol.* 2009 Mar;44(3):209-15. Erratum in: *Pediatr Pulmonol.* 2009 May;44(5):517-8.

Protein-energy malnutrition as the first manifestation of cystic fibrosis in infancy. Fustik S, et al. *Pediatr Int.* 2009 Oct;51(5):678-83.

Statistical limitations of percent ideal body weight as measure for nutritional failure in patients with cystic fibrosis. Hirche TO, et al. *J Cyst Fibros.* 2009 Jul; 8(4):238-44.

Recovery of birth weight z score within 2 years of diagnosis is positively associated with pulmonary status at 6 years of age in children with cystic fibrosis. Lai HJ, et al. *Pediatrics.* 2009 Feb; 123(2): 714-22.

Growth assessment of paediatric patients with CF comparing different auxologic indicators: A multicentre Italian study. Lucidi V, et al. *J Pediatr Gastroenterol Nutr.* 2009 Sep;49(3):335-42.

Association between nutritional status and dietary intake in patients with cystic fibrosis. Simon MI, et al. *J Bras Pneumol.* 2009 Oct;35(10):966-72.

Randomized clinical trial of behavioral intervention and nutrition education to improve caloric intake and weight in children with cystic fibrosis. Stark LJ, et al. *Arch Pediatr Adolesc Med.* 2009 Oct;163(10):915-21.

Growth parameters and calcium homeostasis in cystic fibrosis patients with CFTR I1234V mutation. Wahab AA, et al. *Ann Saudi Med.* 2009 Nov-Dec; 29(6):487-8.

LUNG TRANSPLANT

Prior diabetes mellitus is associated with increased morbidity in cystic fibrosis patients undergoing bilateral lung transplantation: an 'orphan' area? A retrospective case-control study. Bradbury RA, et al. *Intern Med J.* 2009 Jun;39(6): 384-8.

Reversibility of cachexia after bilateral lung transplantation. Habedank D, et al. *Int J Cardiol.* 2009 Mar 20;133(1):46-50.

Obesity and underweight are associated with an increased risk of death after lung transplantation. Lederer DJ, et al. *Am J Respir Crit Care Med.* 2009 Nov 1;180(9):887-95.

Transplant recipients on the edge of the hypocalcemia abyss. Lee P, et al. *J Heart Lung Transplant.* 2009 Jan;28(1):93-5.

Distal intestinal obstruction syndrome (DIOS) in patients with cystic fibrosis after lung transplantation. Morton JR, et al. *J Gastrointest Surg.* 2009 Aug;13(8):1448-53.

RENAL

Proteinuria in cystic fibrosis: a possible correlation between genotype and renal phenotype. Cemlyn-Jones J, et al. J Bras Pneumol. 2009 Jul;35(7):669-75.

Renal dysfunction in cystic fibrosis: is there cause for concern? Soulsby N, et al. Pediatr Pulmonol. 2009 Oct;44(10):947-53. [Review.](#)

Renal involvement in cystic fibrosis: diseases spectrum and clinical relevance. Yahiaoui Y, et al. Clin J Am Soc Nephrol. 2009 May;4(5):921-8.

VITAMINS AND MINERALS

Very high-dose ergocalciferol is effective for correcting vitamin D deficiency in children and young adults with cystic fibrosis. Boas SR, et al. J Cyst Fibros. 2009 Jul;8(4):270-2.

Evaluation of salt supplementation in CF infants. Coates AJ, et al. J Cyst Fibros. 2009 Dec;8(6):382-5.

Vitamin D supplementation for cystic fibrosis. Cochrane Database Ferguson JH, et al. Syst Rev. 2009 Oct 7;(4):CD007298. [Review.](#)

Vitamin D deficiency and chronic lung disease. Gilbert CR, et al. Can Respir J. 2009 May-Jun;16(3):75-80.

Bioavailability of oral vitamin E formulations in adult volunteers and children with chronic cholestasis or cystic fibrosis. Jacquemin E, et al. J Clin Pharm Ther. 2009 Oct;34(5):515-22.

Treatment and prevention of vitamin D insufficiency in cystic fibrosis patients: comparative efficacy of ergocalciferol, cholecalciferol, and UV light. Khazai NB, et al. J Clin Endocrinol Metab. 2009 Jun;94(6):2037-43.

Vitamin D in infants with cystic fibrosis diagnosed by newborn screening. Neville LA, et al. J Paediatr Child Health. 2009 Jan-Feb;45(1-2):36-41.

A 4-month-old boy with acrodermatitis enteropathica-like symptoms. Pekcan S, et al. Eur J Pediatr. 2009 Jan;168(1):119-21.

Hypochloremia and hyponatremia as the initial presentation of cystic fibrosis in three adults. Priou-Guesdon M, et al. Ann Endocrinol (Paris). 2010 Feb;71(1):46-50.

ASSORTED INTERESTING TOPICS

Problems with sleep, eating, and adherence to therapy are common among children with cystic fibrosis. Bayer JK, et al. J Pediatr. 2009 Nov;155(5):759-60.

Adherence to treatment in adolescents with cystic fibrosis: the role of illness perceptions and treatment beliefs. Bucks RS, et al. J Pediatr Psychol. 2009 Sep;34(8):893-902.

Association between nutritional status measurements and pulmonary function in children and adolescents with cystic fibrosis. Chaves CR, et al. J Bras Pneumol. 2009 May;35(5):409-14.

Dental and periodontal health status in children affected by cystic fibrosis in a southern Italian region. Ferrazzano GF, et al. Eur J Paediatr Dent. 2009 Jun;10(2):65-8.

Fat-free mass depletion in cystic fibrosis: Associated with lung disease severity but poorly detected by body mass index. King SJ, et al. Nutrition. 2009 Nov 18. [Epub ahead of print]

Nutrition in cystic fibrosis. Matel JL, et al. Semin Respir Crit Care Med. 2009 Oct;30(5):579-86.

Nutrition management of pediatric patients who have cystic fibrosis. Michel SH, et al. Pediatr Clin North Am. 2009 Oct;56(5):1123-41. [Review.](#)

Nutritional, clinical and socioeconomic profile of patients with cystic fibrosis treated at a referral center in northeastern Brazil. Pinto IC, et al. J Bras Pneumol. 2009 Feb;35(2):137-43.

Prolonged TPN during pregnancy in a cystic fibrosis patient with chronic pancreatitis. Sciaky-Tamir Y, et al. Eur J Obstet Gynecol Reprod Biol. 2009 Mar;143(1):62-3.

Growing old with cystic fibrosis – the characteristics of long-term survivors of cystic fibrosis. Simmonds NJ, et al. Respir Med. 2009 Apr;103(4):629-35.

The long term efficacy of gastrostomy feeding in children with cystic fibrosis on anthropometric markers of nutritional status and pulmonary function. Truby H, et al. Open Respir Med J. 2009 Sep 4;3:112-5.

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