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Title: Early deaths in cystic fibrosis: Analysis of causes and risk factors in a pediatric cohort

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Body: Early diagnosis and proper care improved survival in Cystic Fibrosis. Aim: analysis of causes and risk factors associated with premature deaths in a pediatric CF cohort. Methods: retrospective clinical file analysis of patients followed up during 1999-2011 in a Children's Hospital correlated with level of funding and standards of care. Results: 50 patients diagnosed and followed up, mean age at diagnosis 11 months (90% before age of 1 year). 13/50 patients had meconium ileus (MI), all underwent surgery, 80% complications rate. 20/50 patients died, 11 before 1 year of age; 50% before 2004 (national funding available as source of care). Mean age at diagnosis in deceased patients was 5 months vs.15 months in the still followed up. Overall prevalence of del F508 was 67%; 55% homozygous status. Identified risk factors for early death were: 85% low socioeconomic status and poor understanding of disease, 80% male gender, 75% homozygous del F508, 60% malnutrition at the time of diagnosis. All 6 deceased MI patients had poor outcomes (reinterventions, poor nutrition, early lung involvement). Severe ABPA, liver disease lead to death in 2 patients. Main cause of death was severe pulmonary disease associated with severe malnutrition in some patients. Conclusions: severity of simptoms and rate of disease progression varied widely; and early death was mainly influenced by poor socioeconomic status and presence of del F508 mutation. With current treatment strategies, specialized care, the majority of patients should reach adulthood with good quality of life. This could be achieved in developing countries too, with involvement of more dedicated clinicians and funding directed to organising of care.