

primary studies - published RCT

Cystic fibrosis-related deaths in infancy and the effect of newborn screening.

Study design (if review, criteria of inclusion for studies)

RCT between 1985--1989; intention to treat analysis

Participants

infants, born in Wales and the West Midlands. Eligible children with CF who died in the first 5 years of life were identified from the local pediatricians and from the National UK CF Survey. In all, 230,076 infants were randomized to be screened, while 234,510 were unscreened

Interventions

newborn CF screening by heel-prick immunoreactive trypsin (IRT) measurement or diagnosis by clinical presentation.

Outcome measures

death prevalence, causes of death, median age of diagnosis, clinical presentation

Main results

176 CF children were identified, of whom 7 died in the first 5 years of life, 3 having presented with meconium ileus. Median age of diagnosis in the screened group was 8 weeks. On an intention to treat analysis, all 4 nonmeconium ileus-related deaths occurred in the unscreened group (Fisher's exact test, P

Authors' conclusions

newborn screening has the potential to decrease infant CF deaths, but if it is to be successful, identification and treatment must occur as soon as possible after birth

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See also

Pediatr Pulmonol. 2001 May;31(5):363-6.

Keywords

Gastrointestinal Diseases; Infant; Intestinal Obstruction; Neonatal Screening; Newborn; non pharmacological intervention - diagn; screening; Meconium ileus; diagnostic procedures;