

Cystic fibrosis and meconium ileus: a multicentric study on risk factors for adverse outcome in infancy

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Key words: meconium ileus, atresia, newborn screening

Background. Meconium ileum (MI) is recognized as a risk factor for worse growth in early years in Cystic Fibrosis (CF) CF, as demonstrated by a previous 0-12 month population diagnosed for neonatal screening positivity (FFC 19/2012). We do not yet have epidemiological data or clinical information on early life and on clinical follow-up for Italian CF patients with MI. We suggest that risk factors associated to poor clinical outcomes might be identified in patients with MI, and eventually they might be modifiable. The first aim of the study is to identify risk factors associated to poor clinical outcomes in the first year of life in MI infants. Secondary aim is to describe complications presented in early age.

Methods. Subjects with MI who were born in the years 2009-2015 were enrolled, followed in 13 FC Centers. A database was built for the collection of data: diagnosis of ileum, surgical history, (MI medical or surgical, simple or complex), CF diagnosis data, medical and surgical treatments, anthropometric data and follow up from zero to 12 months. Negative outcome was the failure to grow and/or chronic Pseudomonas infection at 1 year, death. Some variables evaluated as possible risk factors for an adverse outcome are: presence of MI, complicated MI, presence of stoma, duration of the first hospitalization, parenteral nutrition, age at CF specialist visit.

Results. In the study, 85 subjects were enrolled from 13 centers. The study covers 70% of Italian MI cases as reported in the register (85/121). 39 are males.

In 20 subjects (24%) had been prenatal diagnosis of intestinal occlusion and in 11/20 (55%) there was an ileal atresia.

71 (84%) were surgical MI, of which 33 (46%) were complicated.

40/71 (56%) were resected, and in 41 (58%) a stoma was packaged.

In 18/85 (21%) cholestasis was present

In 9 out of 50 (18%) undergone neonatal screening, this was negative

55% (47 subjects) were breast-fed

Full statistical analysis of the risk factors will be presented at the XV Convention FFC

Conclusion. The database has been built so that it could be used for international prospective studies. Since there are no shared recommendations for the management of infant with MI between FC centers and pediatric surgeries, appropriate improvement programs for diagnosis and follow up in the first year of life of MI infants are needed.

References

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PMid:17882440