Objective
To describe the behavioral and emotional profile of children with galactosemia, an inborn error of metabolism. A group of children with phenylketonuria (PKU) was used for comparison. A formal psychological evaluation was conducted for all the children participating in the study. The parents completed two questionnaires, describing the behavior and emotions of their children. There were three major questions in this study: (a) do children with galactosemia have more behavioral and emotional problems? (b) do they have more intelligence or learning difficulties? (c) which is their profile in these characteristics compared to children with PKU?

Material and Methods
A retrospective research was designed, using the psychological test protocols of 13 children with galactosemia and 28 with PKU, who were followed in the Inborn Errors of Metabolism Clinic of Children’s Hospital, Harvard Medical School. The test protocols included formal assessment of the developmental level or IQ of the child, using standardized tests, as well as behavioral/emotional questionnaires completed by the parents. The results were analyzed using ANOVA.

Results
Children with galactosemia had IQ below the normal range, as compared to same-aged peers (from the test’s standardization sample) as well as compared to children with PKU. Children with galactosemia showed difficulty in 7 out of the 17 measures of behavior from the 2 questionnaires.

Discussion
The results of this study show a degree of mental deficiency and the existence of a behavioral/emotional profile that seems to be related specifically to the diagnosis of galactosemia.