## ANALYSIS OF TREATMENT OF PATIENTS WITH MUCOVISCIDOSIS IN MORDOVIA

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**Relevance:** Almost all kinds of cystic fibrosis are genetic in origin. It is a genetic defect that creates fluid build-up in the lungs, which leads to oxygen deprivation and multisystem organ failure. It is rare in most parts of Asia but relatively common in Europe, and in populations of European descent. It is characterized by a defect in the secretion of the extractive glands with a disturbance of the respiratory and gastro-intestinal tract with a severe poor prognosis.

**Objectives:** To study the frequency of occurrence of cystic fibrosis in Mordovia and to inform the audience about the progress in treatment.

**Materials and methods:** Of the 839 thousand inhabitants of Mordovia, only 14 are affected by cystic fibrosis (which is 1.7 per 100 thousand of the population). Of these, 43% are over 18 years old. The average age is  $16.6\pm5$  years. The oldest patient is 49 years old and the youngest is 1.1 years old. Men predominate among the patients — 64%.

**Results:** Progress in detection of cystic fibrosis was achieved due to neonatal screening and genetic investigation at the early stage of development. The proportion of patients diagnosed by neonatal screening was 50%. In Mordovia neonatal screening has been carried out since 2007. Genetic testing was performed in 79% of the total number of patients. Two mutations were identified in 72% of patients, one was identified in 27% of the total number of patients.

**Conclusion:** Analysis of the register of patients with cystic fibrosis in Mordovia showed that the average value of diagnostic indicators, examinations of patients suffering from cystic fibrosis are comparable to the national average.

## Reference

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