European Respiratory Society
Annual Congress 2013

Abstract Number: 2944

Publication Number: P1190

Abstract Group: 7.3. Cystic Fibrosis

Keyword 1: Cystic fibrosis **Keyword 2:** Children **Keyword 3:** Genetics

Title: Evidence incidence of cystic fibrosis: A 30-year study in Krasnoyarsk, Russia

Dr. Vladimir 13490 Chikunov doctorvov@mail.ru , Prof. Dr Natalya 13496 Ilienkova ilenkova1@mail.ru MD and Dr. Tatyana 13544 Elizareva krasmgc@mail.ru . [1] Department of Children Diseases, Krasnoyarsk Medical University, Krasnoyarsk, Russian Federation, 660022 and [2] Laboratory of Genetic, Regional Genetic Center, Krasnoyarsk, Russian Federation .

Body: Monitoring of CF incidence is essential to be able to make reliable predictions for disease management in the future. Methods. This study enrolled CF patients born in Krasnoyarsk region (Siberia, Russia) between January 1st 1981 and December 31st 2012. Patients born before the set up of NBS were retrieved through active diagnosis high-risk groups. In July 2006 NBS for CF was added to the routine screening programme in Krasnoyarsk region. We using the neoIRT-IRT-sweet test protocol. For this protocol, neonates with an neoIRT 70 ng/mL and IRT 40 ng/mL were considered positive, and the standard sweat test was administered to determine CF status. Macroduct or Nanoduct (Wescor, USA) screening systems was use. Patients born since the implementation of NBS were collected by data from genetic centre as well as data from the genetics laboratories of our area. The birth incidence rate of CF (with its 95% confidence interval-95% CI) was determined for the whole study period, by 5-year periods and by year. The incidence rate of a given year was calculated by dividing the number of CF children born over that year by: 1) Prior to the implementation of NBS (1981-July 2006) (n=500, children from group risk), 2) After the implementation of NBS (July 2006-2012) (n=242453). Results. A total of 65 CF children in Krasnoyarsk region between January 1st 1981 and December 31st 2012 were recorded. Half of them (n = 40; 61,5%) were born since the implementation of the NBS program in July 2006. The CFTR genotype could be fully determined in 89,2% of the patients and 56,0% of them were homozygous for the main F508del mutation. The resulting estimate of incidence relative to live births is 1:6061.