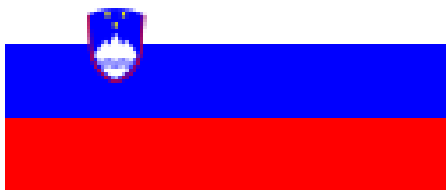




**CENTRE FOR CHILDREN AND ADOLESCENTS,  
UNIVERSITY CLINICAL CENTER, PEDIATRIC CLINIC  
LJUBLJANA, SLOVENIA**

Majda Oštir



# SLOVENIA



## Population

2021: 2.108.977; counting 2002: 1.964.036

**Population density:** 104/km<sup>2</sup>

**Main city:** Ljubljana – 260.000 people

**Official language:** Slovenian, Italian, Hungarian

**Independence from Yugoslavia** was declared d 25.6.1991 and recognized in 1992

**EU entrance** 1 May 2004, €

**GDP 2018** estimate total: usd 75.967 bilion – per capita :USD 36

# SLOVENIA





# 3 CF CENTERS

## 1 PEDIATRIC

## 2 ADULTS



UKC Ljubljana, Pediatric clinic, Department for lung diseases

78

<https://www.ecfs.eu/ecfspr>

27



UKC Ljubljana, Clinical department for pulmonology and lung diseases



8

Clinic for lung diseases and allergy Golnik

# COVID 19 IN OUR CF CENTRE

Prevention activities, CF newsletters, pamphlets about prevention, telephone calls

All outpatient visits (every 3 months) and annual checkups (1 per year) were done.

Data from December 2021

12 (16.6 %) patients recovered from covid 19, all of them unvaccinated before infection

Thirty-five (48,6 %) were fully vaccinated (n=72). Their mean age was  $14 \pm 6.4$  years.

**COVID-19 vaccination in children and adolescents with cystic fibrosis - a single centre experience.** A.Zver<sup>1</sup>, M. Praprotnik<sup>1</sup>, M. Aldeco<sup>1</sup>, D. Lepej<sup>1</sup>, S. Šetina Šmid<sup>1</sup>, J. Rodman Berlot<sup>1</sup>, U. Krivec<sup>1</sup>. *Department for Pulmonology, University Children's Hospital Ljubljana, University Medical Centre Ljubljana, Ljubljana, Slovenia*

# PROPORTION OF CHILDREN (<18 YEARS) AND ADULTS (≥18 YEARS), BY COUNTRY. PATIENTS ALIVE ON 31/12/2019.

Children (<18 years) number (%) 57(50.44)

Adults (≥18 years) number (%) 56(49.56)

Poland	863 (70.11)	368 (29.89)
Portugal	191 (52.47)	173 (47.53)
Romania	229 (94.63)	13 (5.37)
Russian Federation	2526 (74.71)	855 (25.29)
Serbia	134 (66.67)	67 (33.33)
Slovak Republic	134 (41.74)	187 (58.26)
Slovenia	57 (50.44)	56 (49.56)
Spain	1112 (46.26)	1292 (53.74)
Sweden	264 (37.61)	438 (62.39)
Switzerland	427 (42.91)	568 (57.09)
Turkey	1752 (87.95)	240 (12.05)
Ukraine	169 (80.48)	41 (19.52)
United Kingdom	4515 (42.83)	6026 (57.17)
Total	24041 (47.22)	26447 (52.78)

# REVIEW AND ASSESSMENT OF DISEASE INDICATORS IN CHILDREN AND ADOLESCENTS WITH CYSTIC FIBROSIS IN SLOVENIA

Original research article, Slovenian Medical Journal; In press

Ana Drnovšek<sup>1</sup>, Marina Praprotnik<sup>1</sup>, Uroš Krivec<sup>1</sup>, Malena Aldeco<sup>1</sup>, Dušanka Lepej<sup>1</sup>, Saša Šetina Šmid<sup>1</sup>, Aleksandra Zver<sup>1</sup>, Majda Oštir<sup>1</sup>, Katja Seme<sup>2</sup>, Jernej Breclj<sup>3</sup>, Anija Orel<sup>3</sup>, Darja Šmigoc Schweiger<sup>4</sup>, Jasna Rodman Berlot<sup>1</sup>

- (1) *Služba za pljučne bolezni, Pediatrična klinika, Univerzitetni klinični center Ljubljana, Ljubljana, Slovenija*
- (2) *Laboratorij za bakteriološko diagnostiko respiratornih infekcij, Inštitut za mikrobiologijo in imunologijo, Medicinska fakulteta, Univerza v Ljubljani, Ljubljana, Slovenija*
- (3) *Klinični oddelek za gastroenterologijo, hepatologijo in nutricionistiko, Pediatrična klinika, Univerzitetni klinični center Ljubljana, Ljubljana, Slovenija*
- (4) *Klinični oddelek za endokrinologijo, diabetes in presnovne bolezni, Pediatrična klinika, Univerzitetni klinični center Ljubljana, Ljubljana, Slovenija*



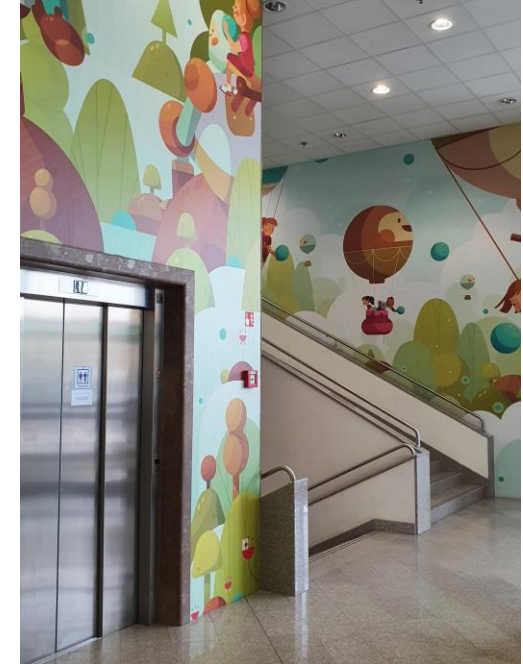
# DEMOGRAPHIC CHARACTERISTICS

78 patients, of whom 56% (44/78) were male patients.

The mean age of the patients was 13.2 years (SD 6.4 years), ranging from 1.2 years to 25.5 years.

The proportion of patients under 18 years of age was 70.5% (55/78).

Two patients after lung transplantation in 2019 and 2020





# GENETICS AND DIAGNOSIS

Mutation of both alleles of the CFTR gene was identified in all patients.

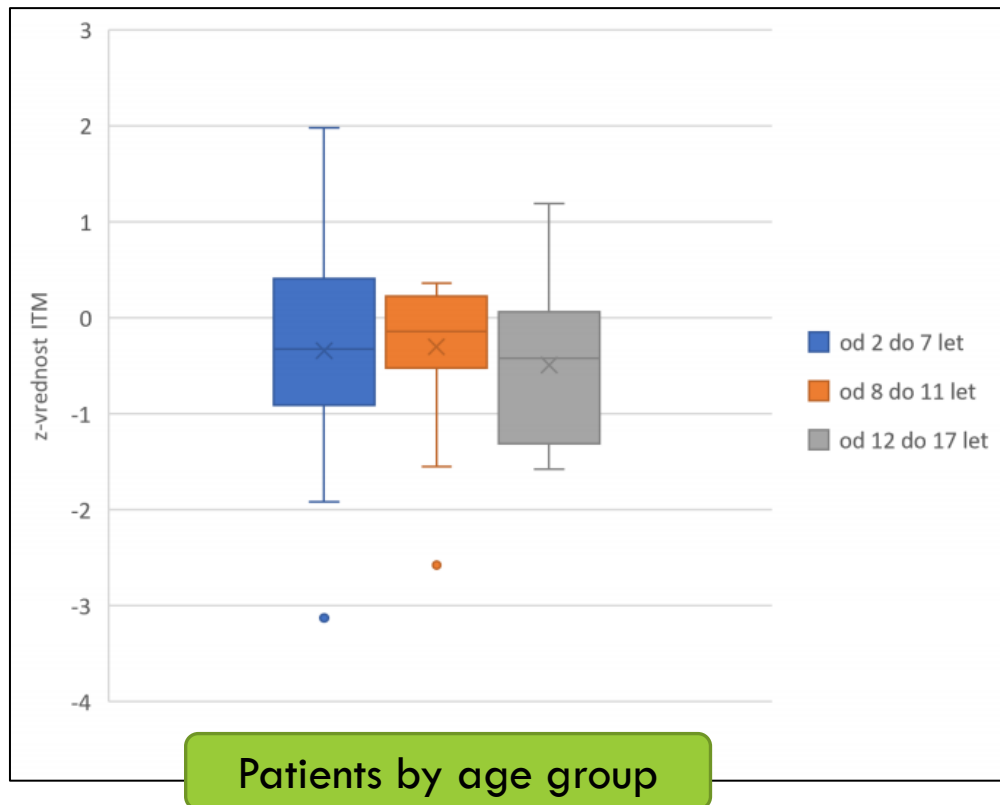
19 different mutations were detected.

The most common F508del, followed by G542X (10.3%), R1162X (9%) and CFTRdel2.3 (3.8%).

As many as 88.5% of patients are carriers of at least one F508del mutation, and in 64.1% of patients, the mutation is present on both alleles.

The median age at diagnosis was 7 months in our patients (IQR 1–24 months)

# NUTRITION

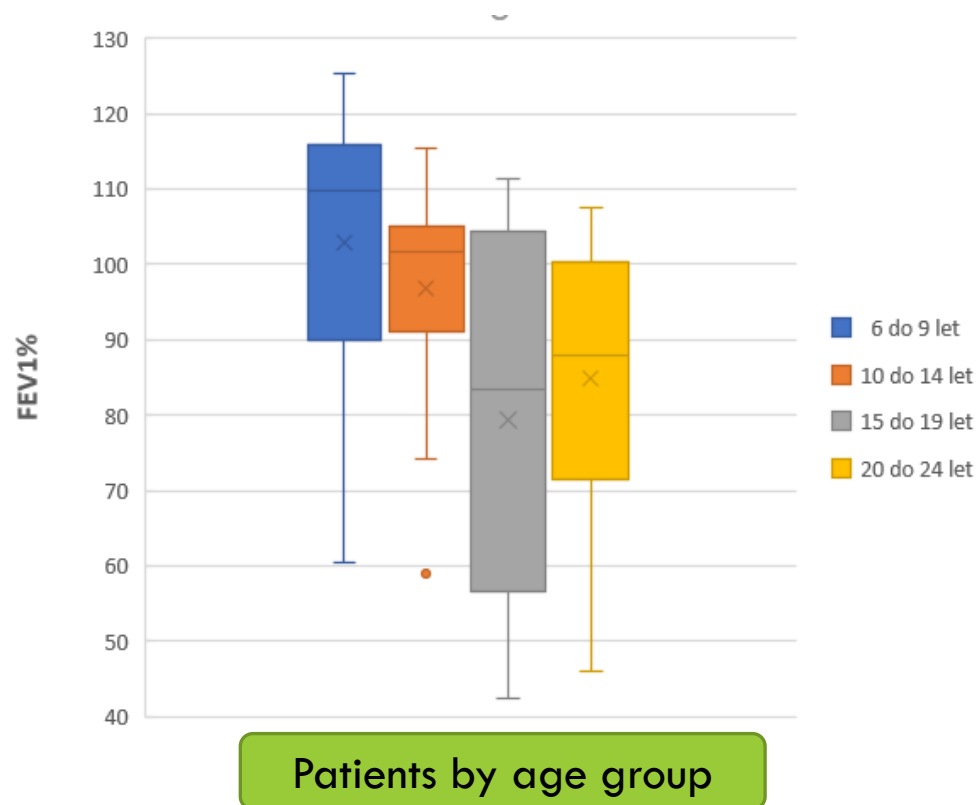


The median z-BMI value was  $-0.2 \text{ kg/m}^2$

In 64.0% (32/50) under 18 years of age, the z-value of BMI was less than  $0.0 \text{ kg/m}^2$  (from  $0.1$  to  $0.4 \text{ kg/m}^2$ )

The mean BMI in patients older than 18 years was  $21.4 \text{ kg/m}^2$

# LUNG FUNCTION



At least one spirometry measurement was performed by 63 patients older than 6 years.

The average FEV1% was 91.3% and the average FVC% was 94.1% (from 102% to 79%)



# MICROBIOLOGY

19.2% (15/78) of all patients had chronic *P. aeruginosa* infection.

Chronic infection was detected in 7.7% (4/52) of patients under 18 years of age.

2 patients with *Mycobacteria abscessus*

4 patients with MRSA

2 patients with *Achromobacter* species





# TREATMENT WITH CFTR MODULATORS IN 2020

## **Lumakaftor / ivacaftor**

Five children (F508 homozygotes) began receiving dual CFTR modulator drug, lumacaftor/ivacaftor (mean age 7.8 years, FEV1% before drug initiation 75.7%).

## **Eleksakaftor / tezakaftor / ivakaftor**

In August 2020, in the program of orphan medicine we started introducing a triple CFTR modulating drug and initially introduced the drug to nine patients with advanced diseases who were carriers of the F508del mutation.

Following the official registration of the drug in November 2020, we continued to introduce the drug to other patients for whom the drug is indicated. By the end of 2020, we introduced the treatment to sixteen patients, which represents 20.5% of all our patients (mean FEV1% before the introduction of the drug 80.7%, LCI before drug administration 15.1).

Following protocol 0, 1m, 3m, 6m, 9m, 12m, 24 m

## Comparison of indicators of diseases of children and adolescents with cystic fibrosis in Slovenia in 2020 with the European average for 2019.

For comparison with other European centers, we used data from the annual report of the European register of cystic fibrosis patients for 2019.

Disease indicator	Slovenija (2020)	EU register (2019)
Age at diagnosis (month), mediana (IQR)	7 (1-24)	4 (1-32)
FEV1 % ( 6-18 let), average (SD)	98,0 % (16,4 %)	90,4 %
BMI z-value (younger from od 18 y), median (IQR)	-0,2 (-0,7-0,2)	-0,2 (-1,0-0,5)
Chronic infections <i>Pseudomona aeruginosa</i> (younger from 18 y), %	7,7 %	12,8 %

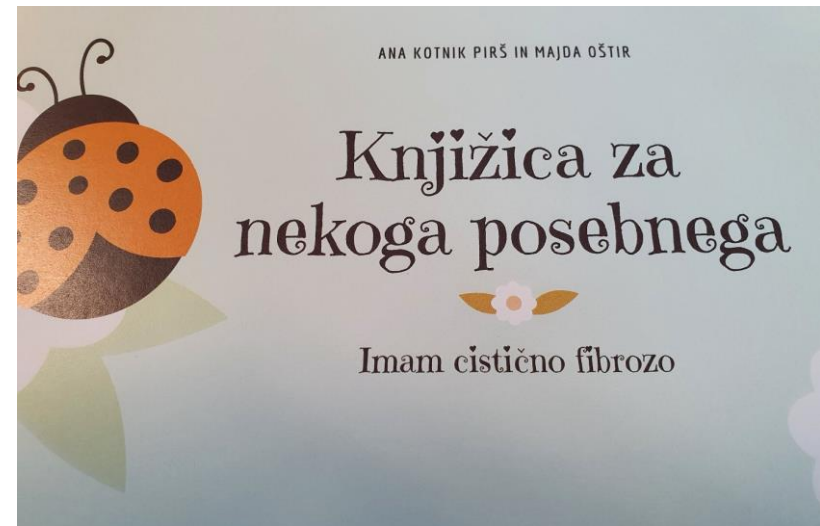
# WORKING WITH AND FOR PATIENTS

- regular monthly CF team meeting
- availability on telephone 8 hours per day (also on email)
- CF newsletter
- booklets for individuals with CF
- Yearly educational day for health professionals working with individuals with CF
- Yearly educational day  
for individuals with CF and their families



[illegible]

Mikrobe lahko dobite in jih širite naprej, ko se dotaknete nečesa, kjer so mikrobi že prisotni, kot sta kljuka ali ograja, in se potem dotaknete svojih oči, nosu ali ust. Umijte si roke z milom in vodo ali jih očistite z gelom za roke na osnovi alkohola. Tudi člane svoje družine in prijatelje spodbujajte, naj imajo čiste roke.





# CONCLUSION

- Waiting for official registration for Eleksakaftor / tezakaftor / ivacaftor for children 6-11 years old (already preparing patients)
- Approved national program for neonatal screening in 2022
- Approved program for improvements in pediatric CF center in 2022
- New edition of Handbook for children and adolescents with cystic fibrosis 2022 – in press
- Challenges in the transition process

