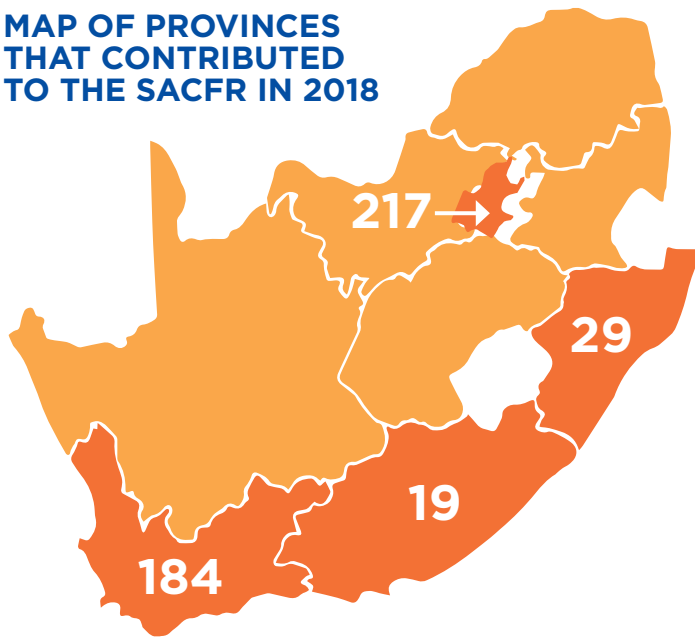




SOUTH AFRICAN
CYSTIC FIBROSIS
REGISTRY
ANNUAL REPORT
SUMMARY 2018



MAP OF PROVINCES THAT CONTRIBUTED TO THE SACFR IN 2018



449*

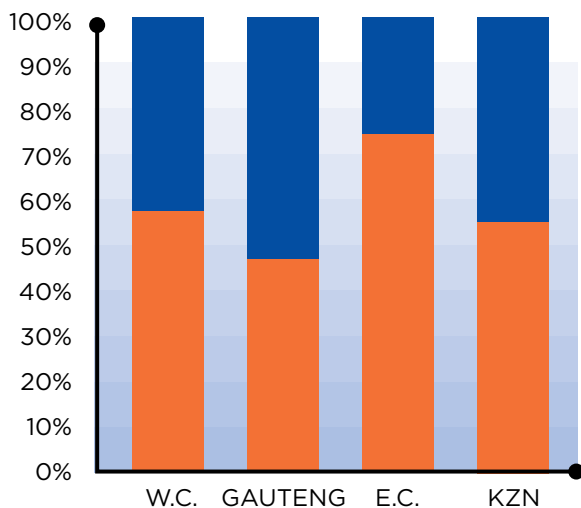
REGISTERED PATIENTS ACROSS 16 CF CENTRES IN 4 PROVINCES

*12 patients were excluded as their diagnosis was unconfirmed

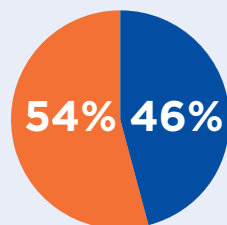
MEDIAN AGE AT DIAGNOSIS

7.7 MONTHS

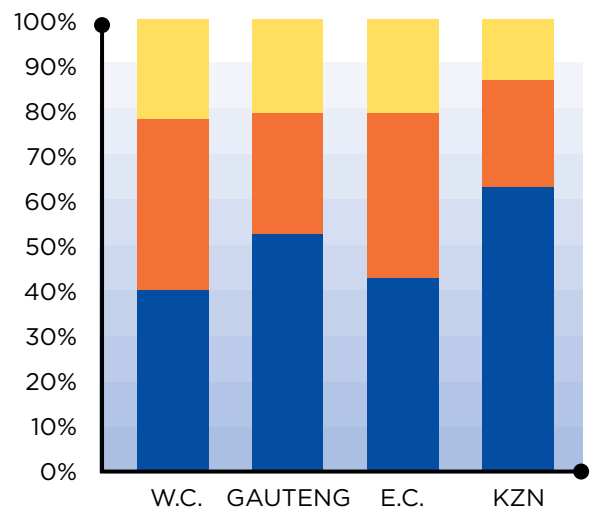
PROPORTION OF CHILDREN AND ADULTS



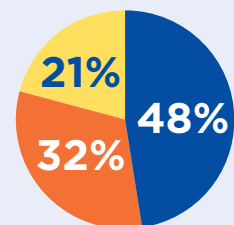
Children
Adults



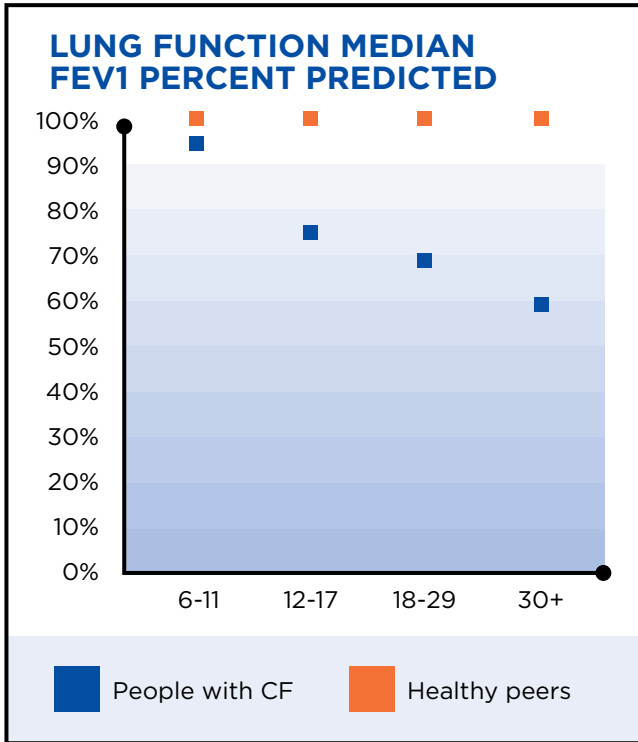
GENETICS



F508del homozygote
F508del heterozygote
No F508del

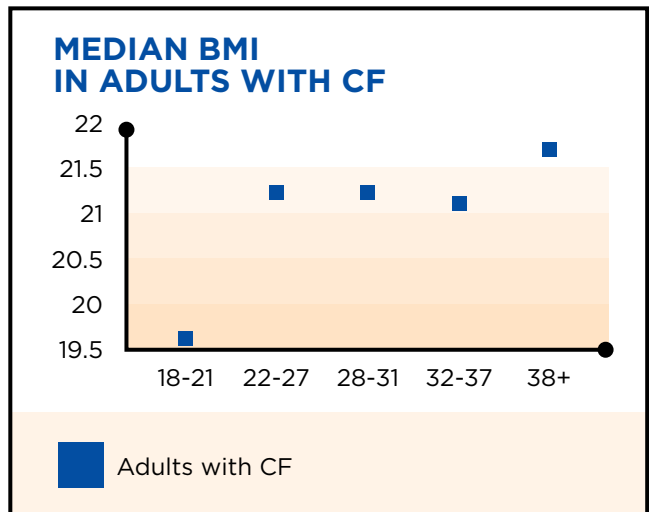
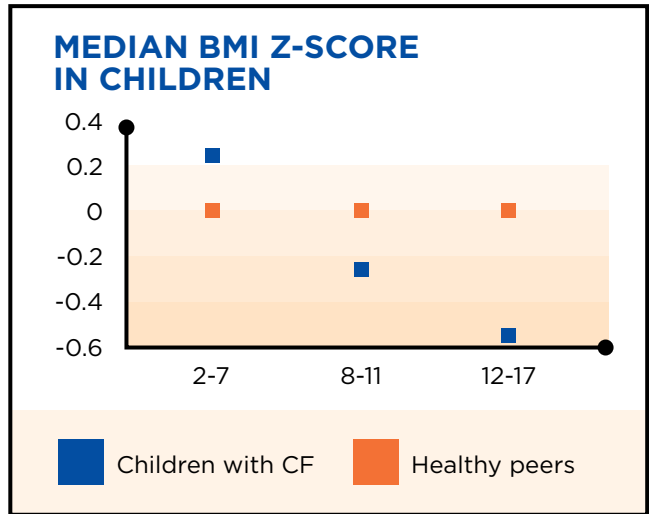


- **F508del** is the most common CF-causing mutation in SA.
- **3120+1G>A** is the second most common CF-causing mutation in SA accounting for 68% of other mutations.
- People with CF have **two CF-causing mutations**, one inherited from the mother and one from the father.
- **Homozygous:** both mutations are the same.
- **Heterozygous:** the two mutations are different.



Excludes 11 patients with previous lung transplants

- **BMI** or **Body Mass Index** is a measure of nutritional status calculated by weight/height².
- **Median** is the middle value.
- A **z-score** indicates how far a value is from the average (mean) value of the reference population status.



CHILDREN (< 18 YEARS)

87% FEV1% MEDIAN (85% MEAN)

ADULTS

65% FEV1% MEDIAN (65% MEAN)

- **FEV1** is a measure of lung function. It is the maximum amount of air that can be forcefully exhaled in the first second after taking a deep breath.
- **FEV1%** is a percentage of the average value for healthy people of the same age, gender and height, which is set at 100%.

- From the age of 8 children with CF have a lower average BMI score than their healthy peers.
- Healthy adult peers have values for BMI between 18 and 25.
- As people with CF get older BMI increases.

26.3%

CHRONICALLY INFECTED WITH PSEUDOMONAS AERUGINOSA IN THEIR LUNGS

34.5% of patients did not have a minimum of 4 sputum tests.

CHRONIC PSEUDOMONAS AERUGINOSA:

CHILDREN: 14%
ADULTS: 42%

27.4%

USE HYPERTONIC SALINE TO HELP CLEAR THICK MUCUS FROM THEIR LUNGS.

50.3%

USE INHALED ANTIBIOTICS TO HELP CLEAR BACTERIAL INFECTIONS IN THEIR LUNGS.

2%

LIVING WITH LUNG TRANSPLANT (n=10)

0.5%

LIVING WITH LIVER TRANSPLANT (n=2)