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Worlds apart: Comparing cystic fibrosis registry cohorts in South Africa and Canada

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Background: Significant differences in epidemiology and outcomes for cystic fibrosis (CF) are expected between low-middle-income and high-income countries, but comparative data are lacking. Comparing CF outcomes between countries may be helpful for planning targeted interventions and advocacy for better care where outcomes are less favorable. This study compared South African and Canadian CF registries' health outcomes and explored factors that may explain differences in outcomes.

Methods: This was a descriptive cross-sectional study of harmonized South African and Canadian CF registries' data from January 1 to December 31, 2018. Demographic, clinical, and outcome variables were compared by calculating standardized mean differences (SMDs) between countries, with a SMD greater than 10 considered clinically important. Genotypes were classified as minimal function (class I-III) or residual function (class IV-VI) CF transmembrane conductance regulator (CFTR) mutations and *Pseudomonas aeruginosa* infection status as *any* in 2018. Outcomes compared were best stable percentage predicted forced expiratory volume in 1 second (FEV_1pp) using ethnic-specific Global Lung Initiative reference standards and body mass index (BMI). Poor nutrition was defined as a BMI z-score of less than −1 in children and a BMI less than 18.5 kg/m^2 in adults. Multivariable regression modeling was performed to adjust for population differences in preselected variables. People taking CFTR modulators and lung transplant recipients were excluded from formal analyses.

Results: Data on 4,049 Canadians and 446 South Africans were analyzed after excluding 585 Canadians taking CFTR modulators. South Africans were younger (median age 15.8 vs 24.1; SMD = 52) and less likely to be male (47.8% vs 54.2%; SMD = 12.5) and Caucasian (70.9% vs 93.3%; SMD = 61.3) than Canadians. Age of diagnosis was similar in both countries, but more children were diagnosed by newborn screening in Canada than South African (n = 550 vs n = 1; SMD = 54.6). Frequency of class I-III CFTR mutations was similar in South Africa (n = 384, 86%) and Canada (n = 3426, 85%); allele frequency of F508del was similar (South African 80% vs Canada 84%). In South Africa, 3120+1G >A (class I) was the second most prevalent mutation (allele frequency 9.9%). *P. aeruginosa* was isolated more frequently in South Africa than Canada (South Africa, n = 192, 43.0% vs Canada, n = 1,458, 36.0%; SMD = 26.3). Three hundred seventy-six (9.3%) Canadians received a lung transplant in 2018 or earlier, compared with 11 (2.5%) in South Africa. FEV_1pp was significantly lower at all ages in South Africa than in Canada, including children aged 6 to 12 (Figure 1), and poor nutrition was significantly more prevalent across all age groups in South Africa than Canada (South Africa, n = 91, 22.5% vs Canada, n = 373, 11.0%; SMD = 31.2, including children aged 0 to 6. After adjusting for age, sex, age at diagnosis, genotype, *P. aeruginosa* infection, and nutrition (lung function model), FEV_1pp was 11% lower (95%CI, −13.35 to −8.21) and poor nutrition was 1.8 times as likely (95%CI, 1.37–2.42) in South Africa than Canada.

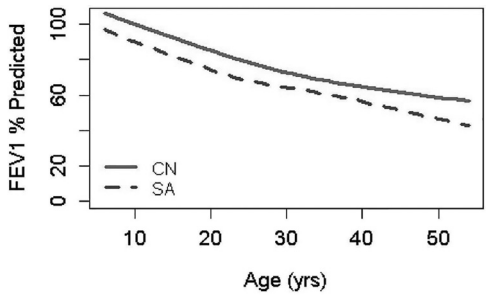

Figure 1. Lung function (percentage predicted forced expiratory volume in 1 second [FEV_1pp]) according to age (≥6) in the South African (SA) and Canadian (CN) cystic fibrosis registry cohorts, 2018

Conclusions: Lung function and markers of nutrition were significantly lower in people with CF in South Africa than in Canada. Nonbiological factors such as lack of newborn screening, early-life factors, poor socioeconomic circumstances, different standards of CF care, and limited access to CFTR modulators and lung transplantation may be important determinants of CF outcomes in South Africa. Analysis to explain disparities in CF outcomes between South Africa and Canada are ongoing.

Acknowledgements: This work is supported by the Cystic Fibrosis Foundation (ZAMPOL19KO), the Harry Crossley Foundation, and the Molly McNeil Family Trust.

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Analysis of survival in cystic fibrosis for a state of Argentina: 2016–2020

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Background: Having a local cystic fibrosis (CF) data registry is essential to learn about the progression of CF in a specific population group. It also helps to understand epidemiological trends in CF mortality and clinical care. The objective of this study was to analyze data from 2016 to 2020 from Buenos Aires Province (population 17 million) retrospectively using a standardized approach to data processing and survival calculations.

Methods: This was a retrospective study. All people with CF registered in the province's reference centers were evaluated with data from the registry. Survival analysis was performed following the Kaplan-Meier methodology. Subjects were considered lost to follow-up (censored) if their last report was more than 2 years before the study completion date. Survival curves differentiated according to sex were assessed.

Results: Of 582 patients (271 female, 311 male), 267 were adults. Probability of survival was 99% for 9.4 years (95% CI, 0.9–11.9 years), 95% for 18.4 years (95% CI, 17.3–23.8 years). At 90% for 24.4 years (95% CI, 21.4–27.5 years), and 80% for 29.4 years (95% CI, 27.5–31.3 years). Median survival was 49.6 years (95% CI, 44.6- years). Average age at death was 25. As reported in the literature, the female survival curve was lower than the male.

Conclusions: This study constitutes the second reporting period describing survival in people with CF from Buenos Aires. The sample size was 28.5% larger than that of the first reporting period. Special emphasis was placed on the improvement in the registry of adult patients, an increase of 60%. Survival values similar to those reported in the previous period were confirmed.