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Jordan

Cystic Fibrosis

National Annual Report 2022

Table of Contents

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List of Figures

List of Tables

FORWARD

In our enduring commitment to the health and wellbeing of every Jordanian, the Ministry of Health, in collaboration with the USAID Health Services Quality Accelerator Activity, takes immense pride and responsibility in presenting the first National Statistical Cystic Fibrosis (CF) Report for Jordan. This pioneering document stands as a testament to our unwavering dedication to understanding, addressing, and ultimately improving the lives of those among us living with cystic fibrosis.

Cystic fibrosis, a life-limiting genetic disorder that significantly affects the respiratory and digestive systems, poses substantial challenges to those diagnosed with the condition, their families, and the healthcare system. Until now, the prevalence, demographic characteristics, and specific healthcare requirements of individuals with CF in Jordan have been poorly understood, primarily due to a lack of comprehensive and centralized data. This inaugural report signifies a monumental step towards changing that narrative. It encompasses critical statistical analyses, from the geographical distribution of CF to healthcare utilization patterns, genetic diversities, and survival rates. Furthermore, it sheds light on the diagnostic paradigms, treatment modalities, and the psychosocial impacts of CF, providing a holistic view of the condition in the Jordanian context.

The implications of this report are multifaceted. For our healthcare professionals, it offers insights that are paramount for enhancing clinical practice, paving the way for personalized medicine, and informing future medical training and research initiatives. For patients and families, it serves as an affirmation of the nation's recognition of their struggles and a commitment to improving their healthcare journey. For policymakers and stakeholders, it provides a data-driven foundation for strategizing, funding, and implementing targeted healthcare policies that address the needs of the CF community.

Moreover, this report underscores the importance of establishing a national electronic CF registry, promoting newborn screening programs, and fostering regional and international collaborations for research and resource sharing. It serves as a direct call for the integration of advanced therapies and innovative care models, emphasizing the need for a multidisciplinary approach to enhance the quality of life for individuals with CF.

As we disseminate this report, we acknowledge that our work does not end here. This document is both a culmination and a beginning — a springboard for continuous action and advocacy. The Ministry of Health is steadfast in its resolve to use the findings from this report to spearhead initiatives that bolster healthcare services, promote equitable access to care, and nurture research in cystic fibrosis.

We extend our deepest gratitude to everyone who contributed to this report and to those who tirelessly serve our citizens affected by cystic fibrosis. Together, we reaffirm our commitment to fostering a healthier Jordan, where every individual has the opportunity to lead a fulfilling life.

Minister of Health

Prof. Feras Hawari

EXECUTIVE SUMMARY

The "2022 National Annual Report for Cystic Fibrosis in Jordan." is the first report providing comprehensive information on patients with a confirmed diagnosis of cystic fibrosis (CF) disease based on the Jordan CF registry data tool. During the reporting period of January through December 2022, 1,784 potential cases of CF were reported from all health sectors in Jordan, of which 385 were confirmed to have the disease based on a review of all reported clinical and historical records for each patient.

The records show that the average age of CF patients was 17 years, with the majority (61%) living in Amman and Irbid. Nearly half of the patients were diagnosed before the age of nine years, with an average age of three years at diagnosis. The gender distribution of CF patients was almost equal, and most patients (97%) were not married. The majority were Jordanians who had health insurance. Forty-seven percent of patients were diagnosed with chloride titration and 18.2% were diagnosed with the conductivity method.

The report provides insights into the most common genetic mutations observed among CF patients in Jordan. Based on the available data the main genetic mutation was Delta F508, offering valuable information for personalized treatment approaches. There was a lack of documented data on genotyping and mutation testing.

Respiratory infections are common among CF patients. About half of the CF cases had no documentation of microbiological testing performed. Of the documented cases, most had bacterial growth, and a few had fungal and bacterial infections. One-third of cases reported hemoptysis. The majority of patients had no lung fibrosis or respiratory failure. While gastrointestinal hepatobiliary manifestations are less common than respiratory manifestations, around 3.1% had gastrointestinal reflux disease, 0.8% had gastrointestinal bleeding, 1.3% had cirrhosis/portal hypertension, 2.6% had pancreatitis, and 1.3% had hepatomegaly.

The average length of CF patients' hospital admission was 16 to 17 days. Around 36% of cases received inhaled antibiotics, while 60% received inhaled bronchodilator, 25% received inhaled mucolytics like hypertonic saline, 35% received inhaled steroids, and only six patients received non-invasive ventilation such as CPAP/BiPAP at home. Similarly, only six patients regularly received chest physiotherapy (CPT) from health professionals or family caregivers. This indicates the need to increase the awareness of patients and families about the importance of CPT. Although CFTR modulators are not accessible in Jordan, three CF patients obtained them from outside the country. The majority of patients received pancreatic enzyme medications. About half of the patients received multivitamins, mainly vitamins A, B, C, D, E, and K, and 16% received nutritional supplements. Only three patients, all infants, received tube feeding.

This report provides valuable insights into the state of CF in Jordan. It serves as a foundation for informed decision-making, resource allocation, and advancements in CF care that can improve the lives of CF patients and contribute to finding a cure for this challenging condition.

INTRODUCTION

Supported by the United States Agency for International Development (USAID), the Health Services Quality Accelerator (HSQA) Activity works with the Government of Jordan to enhance equitable reproductive, maternal, newborn, and child health (RMNCH) outcomes, with a specific focus on disadvantaged populations. This includes improving data management and utilization in the health sector.

The Activity collaborated with the Ministry of Health's (MOH) Non-Communicable Diseases Directorate (NCDD) and a multidisciplinary group of professionals and health care providers to compile and analyze data on all recorded CF patients in Jordan in 2022. This report is the first national CF patients' registry report.

Background

CF is an autosomal recessive genetic disease that affects multiple systems and impacts over 72,000 patients globally.[1] Its prevalence ranges from one in 2,000 to one in 100,000, depending on the country.[2] In the Middle East, the prevalence is estimated at one in 30,000 to 50,000, while recent data shows an incidence of one in 2,000 to 5,800 live births. [3]CF is a life-threatening genetic disease that affects airway epithelial cells, causing the buildup of thick, viscous mucus secretions in various organ systems, including the gastrointestinal, pulmonary, and genitourinary systems. While CF is a multisystem disease, its severity and mortality rate increase due to its respiratory manifestations, such as bronchiectasis.[4]

To better understand the prevalence, impact, and management of CF in Jordan, this report begins by outlining the methodology employed in data collection and analysis, ensuring transparency and accuracy throughout the process. It then explores the demographic characteristics and personal information of CF patients, including age and gender distribution, along with other factors that contribute to the disease's manifestations. The report highlights the diagnostic methods and challenges for CF patients in Jordan, providing an overview of the screening programs, laboratory tests, and genetic analyses utilized for early detection and diagnosis. It also discusses the barriers and limitations faced during data collection, allowing for a better understanding of potential areas for improvement.

The report examines treatment approaches and management strategies available for CF patients in Jordan. It also explores the availability and accessibility of medications, therapies, and multidisciplinary care, aiming to identify gaps and areas where additional support and resources may be required.

The report concludes with a summary of key findings, emphasizing the importance of early detection, access to comprehensive care, and ongoing research and recommendations in the field of CF. It calls for collaborative efforts between healthcare providers, policymakers, and patient support groups to improve awareness, education, and support systems for individuals living with CF in Jordan.

Overall, this report is a vital resource for healthcare professionals, researchers, policymakers, and individuals affected by CF in Jordan. It aims to facilitate evidence-based decision-making, promote equitable healthcare access, and ultimately contribute to better outcomes and improved quality of life for those living with CF in the country.

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1 Jackson AD, Goss CH. Epidemiology of CF: how registries can be used to advance our understanding of the CF population. J Cyst Fibros. 2018 May;17(3):297-305. Available from https://pubmed.ncbi.nlm.nih.gov/29275954/ doi: 10.1016/j.jcf.2017.11.013

2 Konstan MW, Pasta DJ, VanDevanter DR, Wagener JS, Morgan WJ. Epidemiologic study of cystic fibrosis: 25 years of observational research. Pediatr Pulm. 2021 May;56(5):823-36. Available from: https://pubmed.ncbi.nlm.nih.gov/33434406/ doi: 10.1002/ppul.25248

3 Banjar H, Angyalosi G. The road for survival improvement of cystic fibrosis patients in Arab countries. Int J Pediatr Adolesc Med. 2015 Jun;2(2):47-58. Available from: https://pubmed.ncbi.nlm.nih.gov/30805437/ doi: 10.1016/j.ijpam.2015.05.006

4 Karrar HR, Noun MI, Alanazi AAG, Alharbi SE, Almutairi FN, Alhammad HAA, Sadeeg AAH, Alsheikh MAA, Alshaikh WM, Alyahya MY, Makki WAS, Alhazmi HM, Almutairi AS, Alhendi RSA. Cystic fibrosis: a review article. World Family Medicine Journal [An Overview of Biological Warfare and SARS-CoV-2 as a Potential Biological Agent]. 2022 Mar;20(3):58-63. Available from: http://www.mejfm.com/March%202022/Cystic%20Fibrosis.pdf doi: 10.5742/MEWFM.2022.9525014

STEPS IN DEVELOPING THE CF REPORT

STEPS IN DEVELOPING THE CF REPORT

The following steps were taken to develop this first national CF statistical report for Jordan:

1. Objectives defined: The national CF registry development steering committee conducted a series of meetings to clearly articulate the goals and objectives of establishing a national CF registry for Jordan. Objectives included determining the prevalence of the disease, understanding the demographics of the affected population, identifying treatment patterns, and assessing the overall burden of cystic fibrosis on the healthcare system in Jordan.
2. Data sources identified: The committee collaborated with hospitals and healthcare facilities across Jordan to identify all locations where patients diagnosed with CF receive treatment. This included specialized CF clinics, pediatric departments, and respiratory care units across the kingdom.
3. Data collection instruments developed: A standardized CF registry data collection tool was developed to gather information from both medical records and patient interviews. The tool was designed to match the international CF foundation registry data tools. It was contextualized to the Jordanian health system to capture key data points such as patient demographics, medical history, diagnostic methods, treatment modalities, and outcomes. The tool was piloted to ensure it was consistent and easily understandable by the participating healthcare professionals and consultants.
4. Ethical approval obtained: Ethical approval was obtained from patients/parents in compliance with data protection and patient privacy regulations using the consent form attached as Annex A.
5. Data collectors trained: Data collectors were trained, including healthcare professionals responsible for data abstraction from medical records, and interviewers to engage with patients. Data collectors were provided comprehensive guidance on the data collection instruments, ethical considerations, and proper documentation techniques on the adjusted digitalized tool. Live examples were provided on the scenarios or challenges that might be faced during the data collection process.
6. Meeting held with the International Cystic Fibrosis Foundation: A virtual learning session was held on November 14, 2022, with the International Cystic Fibrosis Foundation to learn from their experience in establishing CF patient registries in different countries worldwide.
7. Data collected from medical records: The trained data collectors (one pediatric physician, two general practitioners, and one critical care nurse) were assigned to abstract relevant data from medical records of diagnosed CF patients across all hospitals and clinics in Jordan. This included information on diagnosis, clinical characteristics, treatment history, co-morbidities, and any other variables as per the objectives defined earlier and based on the registry data collection tool.
8. Patients interviewed: The data collection team scheduled interviews with diagnosed CF patients or their parents, if they were under 18 years old, to gather additional information. These interviews helped validate and supplement the data collected from medical records using the standardized interview questionnaire to inquire about patient experiences, treatment adherence, quality of life, and any other relevant factors.
9. Data validated and quality checked: The team implemented a robust data validation process to ensure the accuracy and consistency of the collected information. The data obtained from medical records was cross-checked with the patient interview responses to identify any discrepancies or missing data. Data cleaning and quality control measures were performed to ensure the reliability of the dataset.
10. Data analysed and report prepared: Statistical analysis was conducted using Stata to generate meaningful insights. A comprehensive report was prepared, including tables, figures, and a summary of the findings.

The MOH, in collaboration with the national CF registry development steering committee, will share the national statistical report with relevant stakeholders, including healthcare professionals, policymakers, patient advocacy groups, and researchers. They will also seek to publish the report in scientific journals and make it publicly available to raise awareness about cystic fibrosis and facilitate evidence-based decision-making in healthcare planning and resource allocation.

Data Management

At the outset, a letter was sent to all hospitals in all health sectors in Jordan to request a list of the CF patients treated in each facility.

Figure 1: Letters issued from the MOH to all health sectors in Jordan to share lists of CF patients

A review of records, as shown in Figure 2 below, initially identified a total of 1,784 patients who were diagnosed as CF.

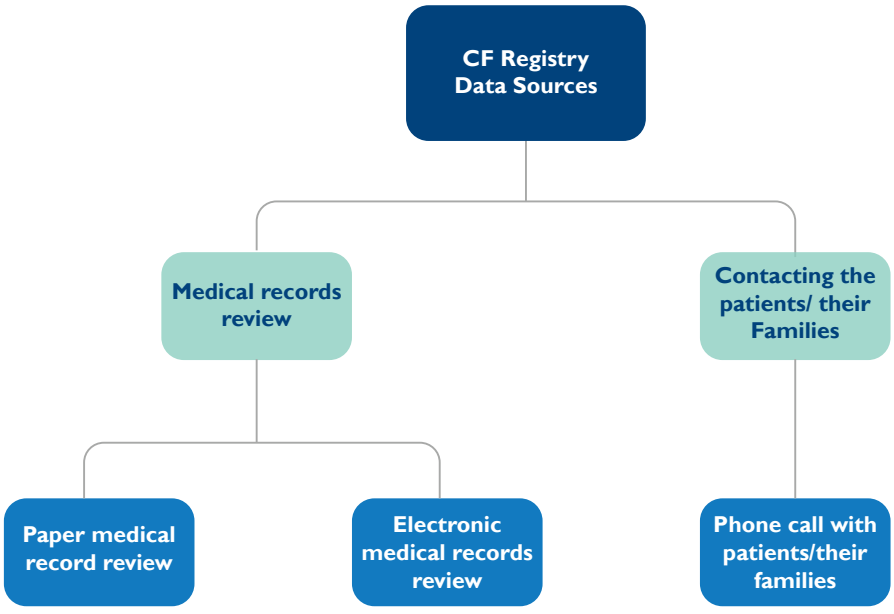


Figure 2: CF registry data collection sources

However, analysis of these cases found that 920 were duplicates based on national or personal ID numbers. This left 864 cases for auditing their medical records to confirm clinical manifestations. The audit by qualified and trained physicians resulted in a total of 385 confirmed cases of CF.

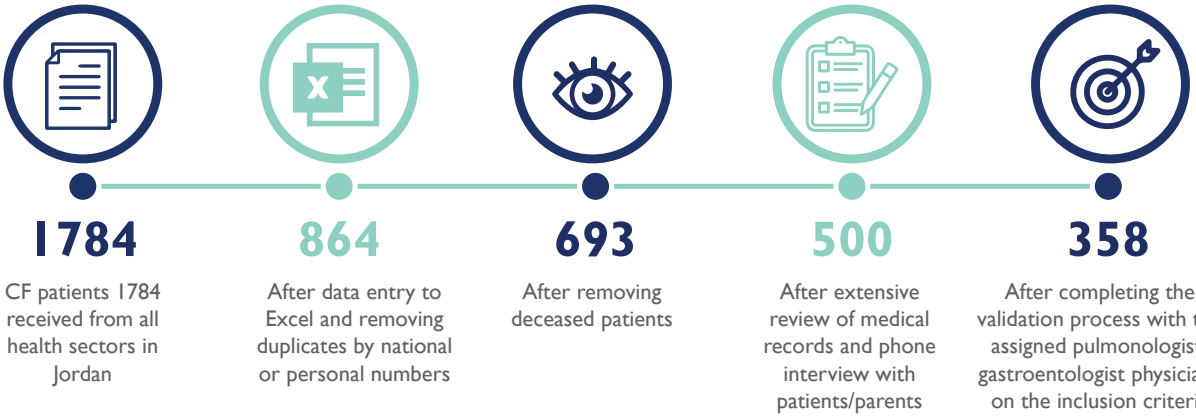
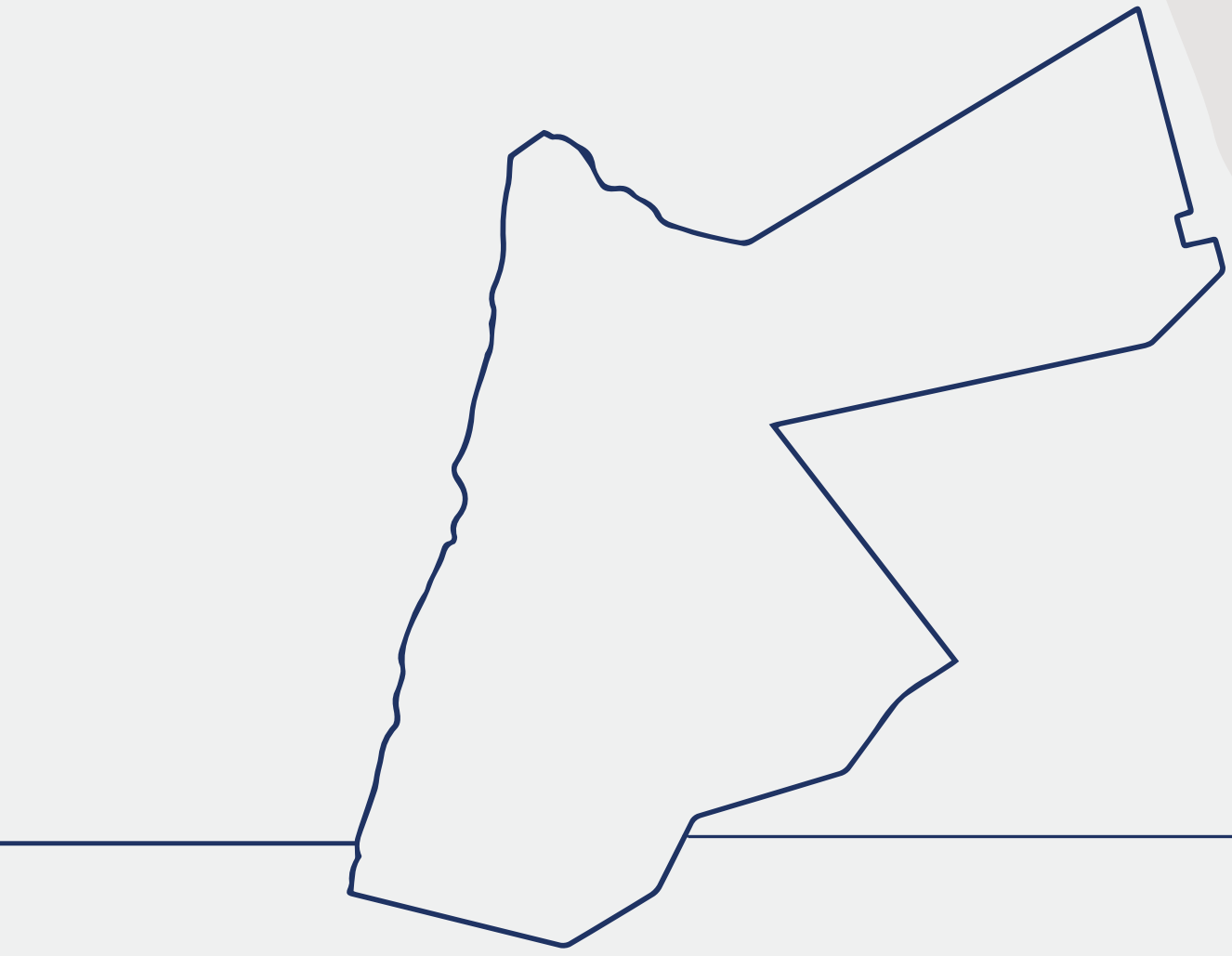


Figure 3: Validation Process for Confirmed CF Cases

Distribution of Cystic Fibrosis Cases in Jordan According to Demographic Variables



RESULTS

This report charts the trends in confirmed, live CF patients in Jordan, examining various dimensions, including patient age, gender, genetic mutations, lung function, nutrition, co-morbidities, treatments, and survival. The findings aim to shed light on how these factors interplay and affect the overall health and lifespan of those living with CF.

Distribution of Cystic Fibrosis Cases in Jordan According to Demographic Variables

Distribution by Governorate of residence

As shown in Table 1, the majority of CF patients reside in the governorates of Amman (35.3%) and Irbid (26%). The remaining patients are distributed across various governorates, except Maan, where no patients were reported during this period. In 13.3% of cases, no information was available on the governorate of residences.

Distribution by age

58.6% of the CF patients were below the age of 20 in 2022, and 80.9% were under the age of 30, highlighting the prevalence of the condition among younger age groups.

The mean age of the identified patients was 17 years, and the minimum age recorded during this period was six months. The maximum age reported was 52 years, indicating that there are cases of cystic fibrosis among older individuals as well.

Distribution by gender

There was an almost equal distribution of CF patients between females and males.

Distribution by marital status

An overwhelming majority of the patients, around 99%, were unmarried.

Distribution by educational level

53.5% of patients were found to have completed secondary education or lower. However, around 41% of the cases did not report their educational level.

Distribution by nationality

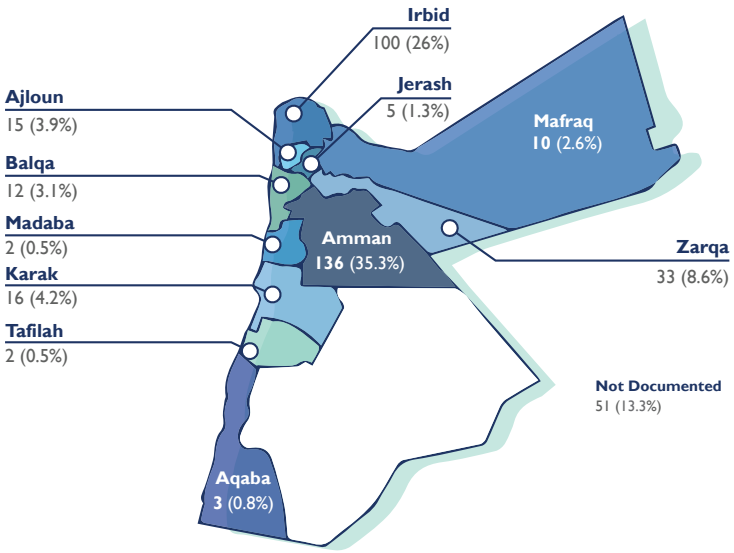
The majority of cases were found among Jordanian individuals, with only six cases reported among Syrian nationals and four among other nationalities.

Distribution by health insurance coverage

Virtually all (97%) of patients had health insurance coverage in the public sector.

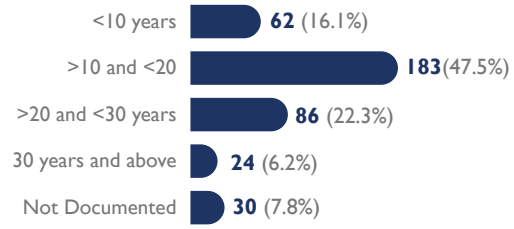
Figure 4: Distribution of Cystic Fibrosis Cases in Jordan According to Background Variables

Governorate

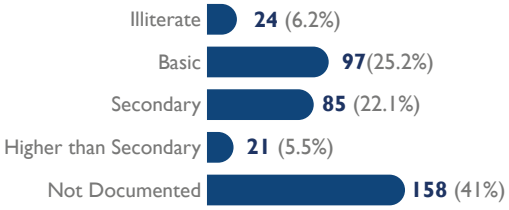


Note: Numbers represent the count of cases associated with cystic fibrosis

Current Age Groups

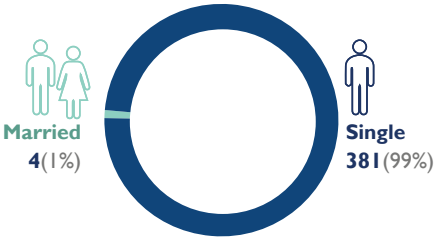


Educational Level

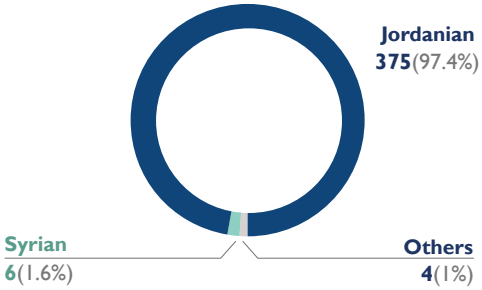


Sex

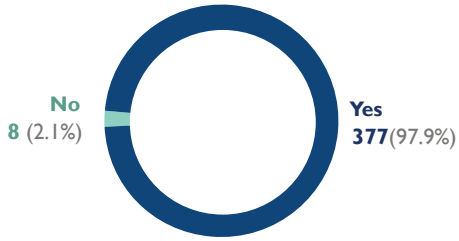
Marital Status



Nationality



Health Insurance



2

Cystic Fibrosis Diagnosis

Cystic Fibrosis Diagnosis

CF is typically diagnosed through a combination of clinical evaluation, genetic testing, and specialized diagnostic tests. For clinical assessment, a healthcare professional examines symptoms such as persistent cough, frequent lung infections, poor growth, and digestive issues. Genetic testing is conducted to identify specific mutations in the CFTR gene responsible for CF. Sweat chloride testing is a key diagnostic test, measuring the chloride concentration in sweat, which is elevated in CF patients. Other tests, including lung function tests and chest X-rays, help evaluate lung health and detect respiratory complications. Major manifestations of CF include chronic lung infections leading to respiratory difficulties, pancreatic insufficiency leading to malabsorption and malnutrition, salty-tasting skin, infertility in males, and possible complications affecting the liver, sinuses, and bones. Early diagnosis and timely intervention are crucial to managing CF effectively and improving the overall quality of life for individuals living with this genetic disorder.

This section, including Table 2 below, presents the distribution of CF cases in Jordan according to diagnosis-related variables. Many variables have missing or undocumented data, a situation that could be improved with a compelling national electronic CF registry platform which would lead to a better understanding of the disease context in Jordan.

Distribution by age at diagnosis

Approximately half of the patients were diagnosed before the age of nine, with one-fourth of them being diagnosed before the age of one year and a small proportion (4.2%) diagnosed at birth. The average age at diagnosis was three years, ranging from a minimum of zero (diagnosis at birth) to a maximum of 33 years.

None of the CF patients were diagnosed during the antenatal period. In fact, CF is not included in the national antenatal screening program.

Distribution by diagnostic tests

The sweat chloride test is a widely used diagnostic tool for identifying CF disease. This test measures the concentration of chloride ions in sweat, as individuals with CF have elevated chloride levels due to malfunctioning chloride channels in their sweat glands. The test involves applying a chemical stimulant, such as pilocarpine, to the skin, which triggers sweating. The sweat is then collected and analyzed to determine the chloride concentration. A chloride concentration above a certain threshold (typically 60 milliequivalents per liter) indicates CF. This non-invasive and relatively simple procedure has proven to be highly effective in diagnosing CF and plays a vital role in confirming the presence of the disease.[5]

It is important to note that the sweat chloride test should be interpreted in conjunction with other diagnostic evaluations, including genetic testing and clinical assessment, to ensure accurate diagnosis and appropriate management of CF.[6] Chloride titration and conductivity are two different methods used in diagnosing CF by measuring chloride levels in sweat.

The majority of patients had a clinical manifestation that matched with the CF disease clinical picture. Most patients were not diagnosed by Meconium Ileus. Around two-thirds of patients (65%) had a value for sweat chloride reported in their medical record. The average sweat chloride test was 95 milliequivalents per liter. The lowest reported value of sweat chloride was 45 milliequivalents per liter, while the highest value was 195 milliequivalents per liter.

Around 47% of patients were diagnosed with chloride titration, and 18.2% were diagnosed with conductivity.

5 Mayo Clinic. (2021). Cystic Fibrosis: Diagnosis & Treatment. Retrieved from https://www.mayoclinic.org/diseases-conditions/cystic-fibrosis/diagnosis-treatment/drc-20353716

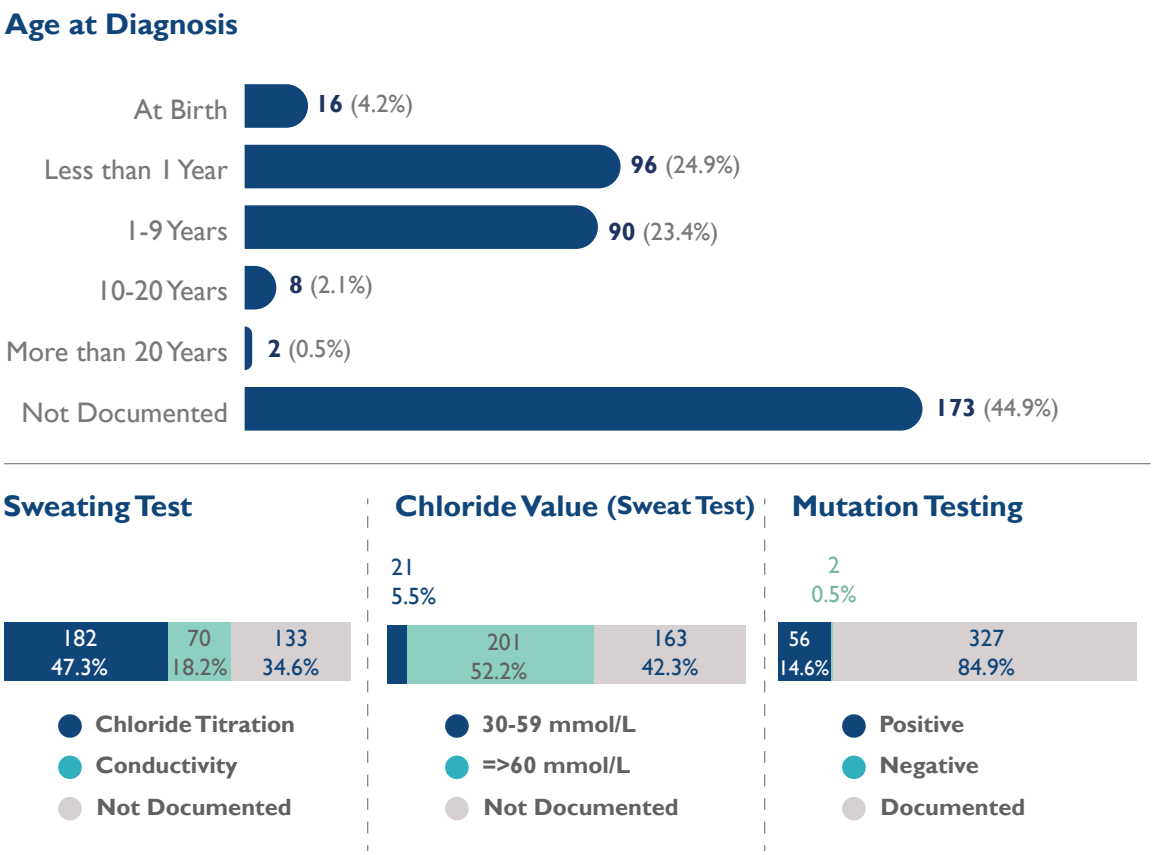
6 Citation: Boyle, M. P. (2014). Diagnosis and management of cystic fibrosis. Clinics in chest medicine, 35(2), 155-164.

Distribution by genotyping and mutation testing

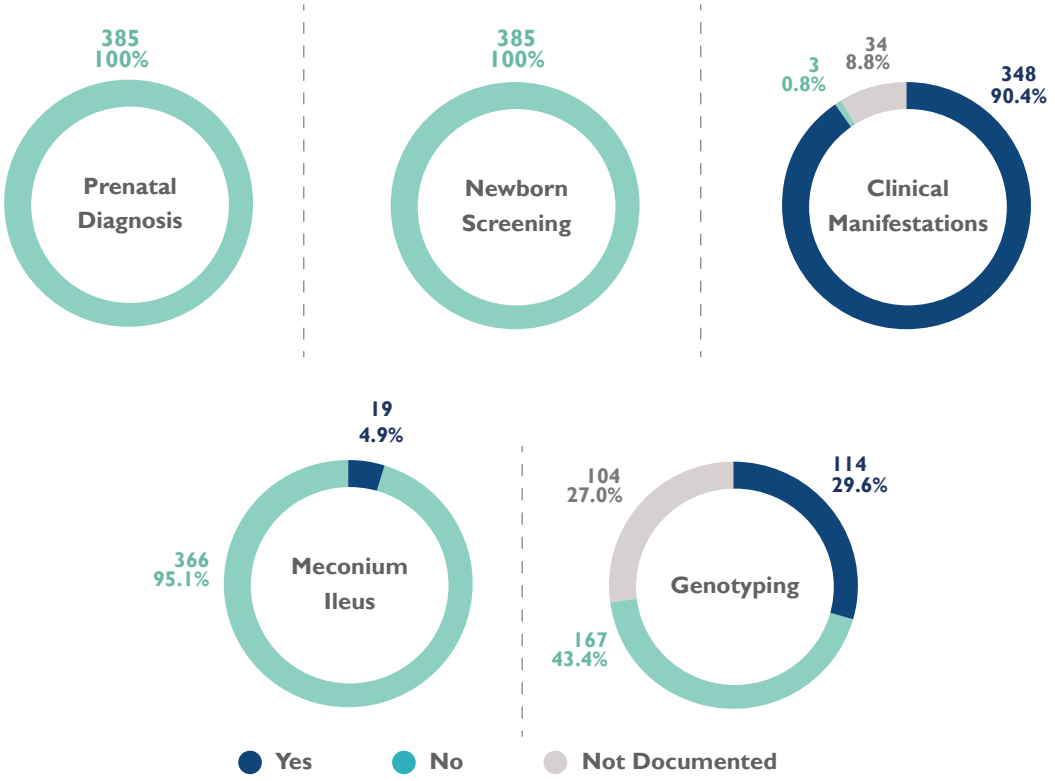
The results of testing conducted within the past five years shed light on the genotyping and mutation testing status of CF patients. Findings revealed that 30% of the patients underwent genotyping, 43% did not; and the remaining patients lacked documented data.A significant majority (85%) of patients had no documented mutation testing results. Among the 58 patients who underwent a documented mutation test, almost all (56) were positive. Different mutations were found, with the most common in Jordan being delta F 508 (see Annex C).

These findings highlight gaps and limitations in CF patients' genotyping and mutation testing documentation. Accurate and comprehensive genotyping and mutation testing are crucial for precise diagnosis, prognosis, and personalized treatment approaches. It is imperative to address the lack of genotyping and mutation testing data and ensure proper documentation to facilitate improved management and care for CF patients.[7]

Figure 5: Distribution of Cystic Fibrosis Cases in Jordan According to Diagnosis Related Variables



7 Burgel, P. R., Bellis, G., Olesen, H.V., Viviani, L., Zolin, A., Blasi, F., ... & European Cystic Fibrosis Society Clinical Trial Network (ECFS-CTN) Study Group. (2020). Future trends in cystic fibrosis demography in 34 European countries. European respiratory journal, 55(6), 1901851.



Note: Numbers represent the count of cases associated with cystic fibrosis

3

Health and Functioning: Respiratory Infections

Health and Functioning: Respiratory Infections

Table 3 below shows the distribution of CF in Jordan according to respiratory variables.

Distribution by annual X-Ray

Based on the available literature, annual chest X-rays are essential for CF patients as they provide valuable information on pulmonary health, aid in the early detection of complications, monitor treatment response, and guide management decisions. Regular imaging helps ensure proactive and comprehensive care for CF patients, leading to better disease management and improved quality of life[8].

The average number of X-rays reported for patients was 2.23 annually, with a minimum of no x-ray done during 2022 to a maximum of 12 X-rays done during 2022. Around 29% of patients had one to three x-rays in 2022, and 10% had four to six.

Distribution by microbiology testing

Microbiology testing is a vital component in the comprehensive care of CF patients. The necessity of regular microbiology testing is well-supported by research, as it allows for the identification and monitoring of specific pathogens commonly associated with CF lung infections. A study published in the Journal of Cystic Fibrosis in 2020 emphasized the importance of microbiology testing in CF management, highlighting its role in guiding targeted antibiotic therapies and facilitating timely intervention in the event of exacerbations or changes in microbial profiles. Regular microbiology testing enables healthcare professionals to tailor treatment regimens, prevent the development of antibiotic resistance, and improve overall outcomes for CF patients[9].

About half of the CF cases had no documentation of microbiological testing performed. Among those who were tested, most had bacterial growth, and a few had both fungal and bacterial infections.

Distribution by hemoptysis

Assessing hemoptysis is crucial in the management of CF patients. |Hemoptysis refers to significant coughing up of blood and can indicate underlying pulmonary complications such as bronchiectasis or lung infection. It is important to promptly evaluate and address major hemoptysis to prevent further complications and provide appropriate interventions. A study published in the Journal of Cystic Fibrosis in 2020 emphasized the significance of assessing major hemoptysis in CF patients, highlighting the need for immediate medical attention, including respiratory support, bronchial artery embolization, or surgical interventions, depending on the severity and cause of the bleeding[10]. Early identification and management of hemoptysis is essential for optimizing outcomes and improving the overall care of CF patients.

One-third of the cases reported hemoptysis, while 38% reported no hemoptysis, and the rest had no documentation of hemoptysis.

8 Burgel, P. R., Bellis, G., Olesen, H. V., Viviani, L., Zolin, A., Blasi, F., ... & European Cystic Fibrosis Society Clinical Trial Network (ECFS-CTN) Study Group. (2020). Future trends in cystic fibrosis demography in 34 European countries. European Respiratory Journal, 55(6), 1901851.

9 (Cystic Fibrosis Foundation, 2020).

10 Mingora, C. M., & Flume, P. A. (2021). Pulmonary complications in cystic fibrosis: past, present, and future: adult cystic fibrosis series. Chest, 160(4), 1232-1240.

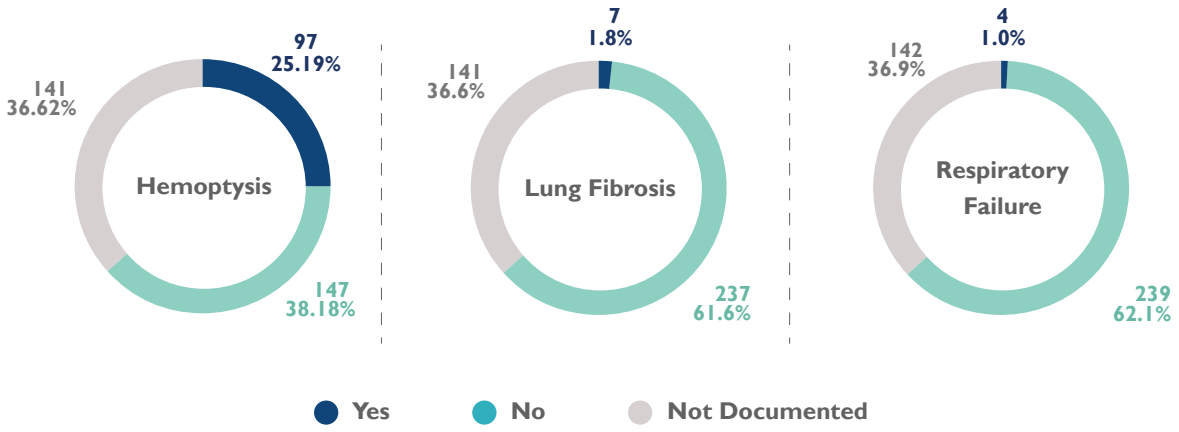
Distribution by lung fibrosis and respiratory failure

Lung fibrosis and respiratory failure are complications that significantly impact lung function and overall prognosis in CF. Regular evaluation of lung fibrosis, typically through imaging techniques like high-resolution computed tomography (HRCT), allows for the early detection and monitoring of structural changes in the lungs. This aids in guiding treatment decisions, such as initiating targeted therapies and interventions to slow disease progression. Furthermore, assessing respiratory failure, typically measured through pulmonary function tests (PFTs), is essential in determining the severity of lung impairment and the need for additional respiratory support or lung transplantation. A comprehensive approach that includes regular lung fibrosis and respiratory failure assessment is crucial for optimizing CF patient care[11].

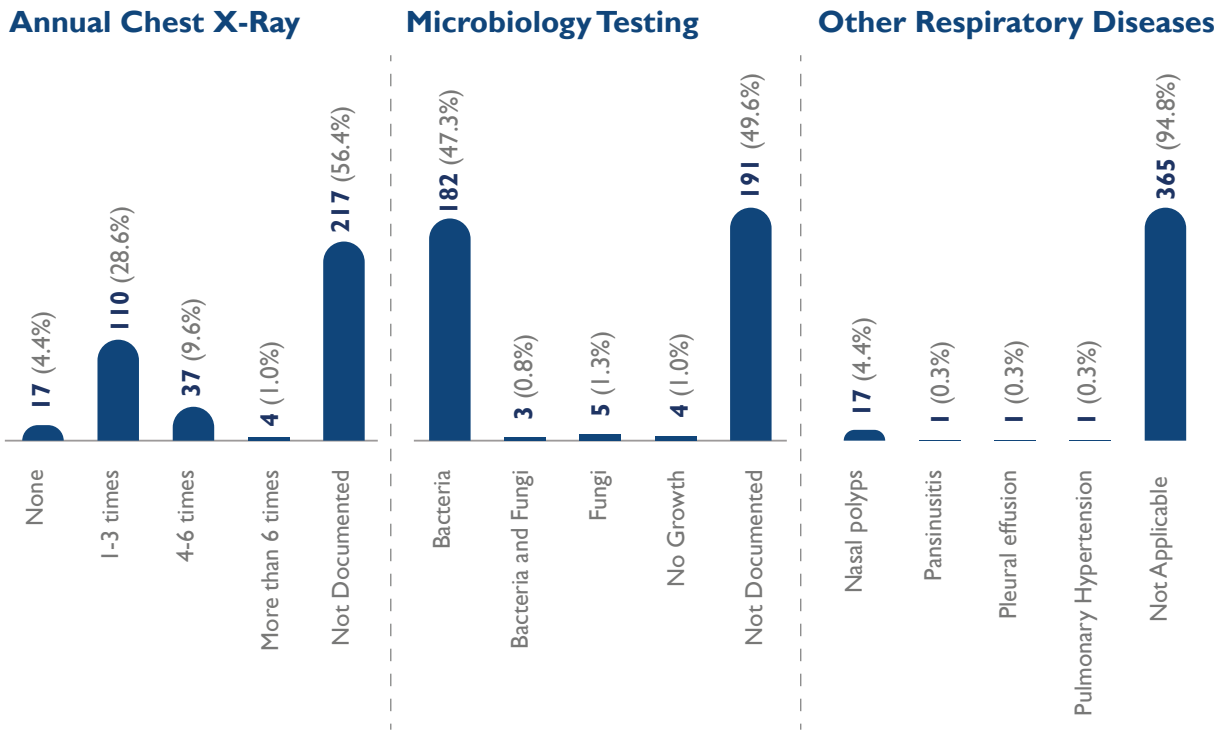
The majority of patients had no lung fibrosis and no respiratory failure. Just one percent reported respiratory failure. However, the data was not documented in around one-third of cases, so the picture needs to be completed. Distribution by another respiratory disease

Other respiratory diseases reported included 14 cases were nasal polyps and one case each of pansinusitis, pleural effusion, and pulmonary hypertension.

Figure 6: Distribution of Cystic Fibrosis Cases in Jordan According to Respiratory Variables

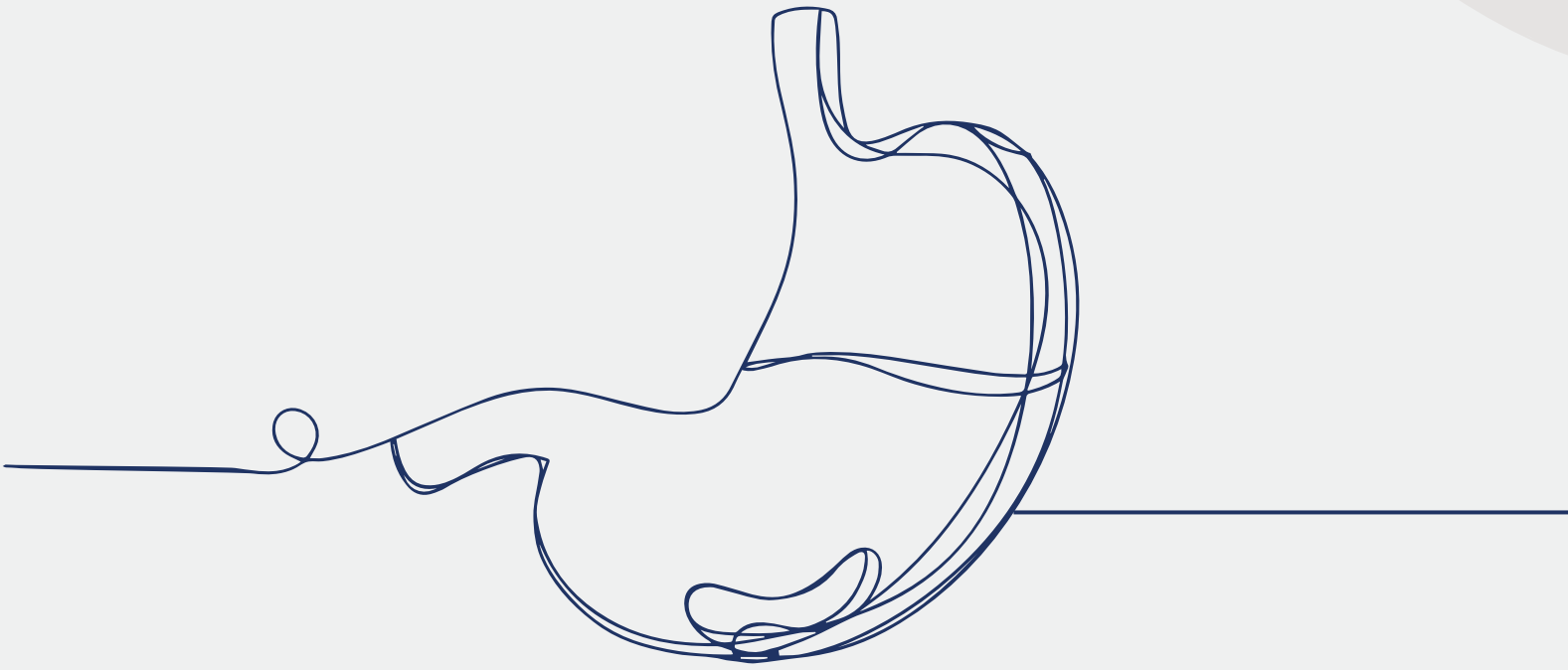


11 Rosenfeld, M., Cunningham, S., Harris, W. T., Lapey, A., Regelmann, W. E., Sawicki, G. S., ... & Konstan, M. W. (2019). An open-label extension study of ivacaftor in children with CF and a CFTR gating mutation initiating treatment at age 2–5 years (KLIMB). Journal of Cystic Fibrosis, 18(6), 838-843.


Annual Chest X-Ray
17 (4.4%)
None
110 (28.6%)
1-3 times
37 (9.6%)
4-6 times
4 (1.0%)
More than 6 times
217 (56.4%)
Not Documented
Microbiology Testing
182 (47.3%)
Bacteria
3 (0.8%)
Bacteria and Fungi
5 (1.3%)
Fungi
4 (1.0%)
No Growth
191 (49.6%)
Not Documented
Other Respiratory Diseases
17 (4.4%)
Nasal polyps
1 (0.3%)
Pansinusitis
1 (0.3%)
Pleural effusion
1 (0.3%)
Pulmonary Hypertension
365 (94.8%)
Not Applicable

4

Cystic Fibrosis and Gastrointestinal/Hepatobiliary Manifestations



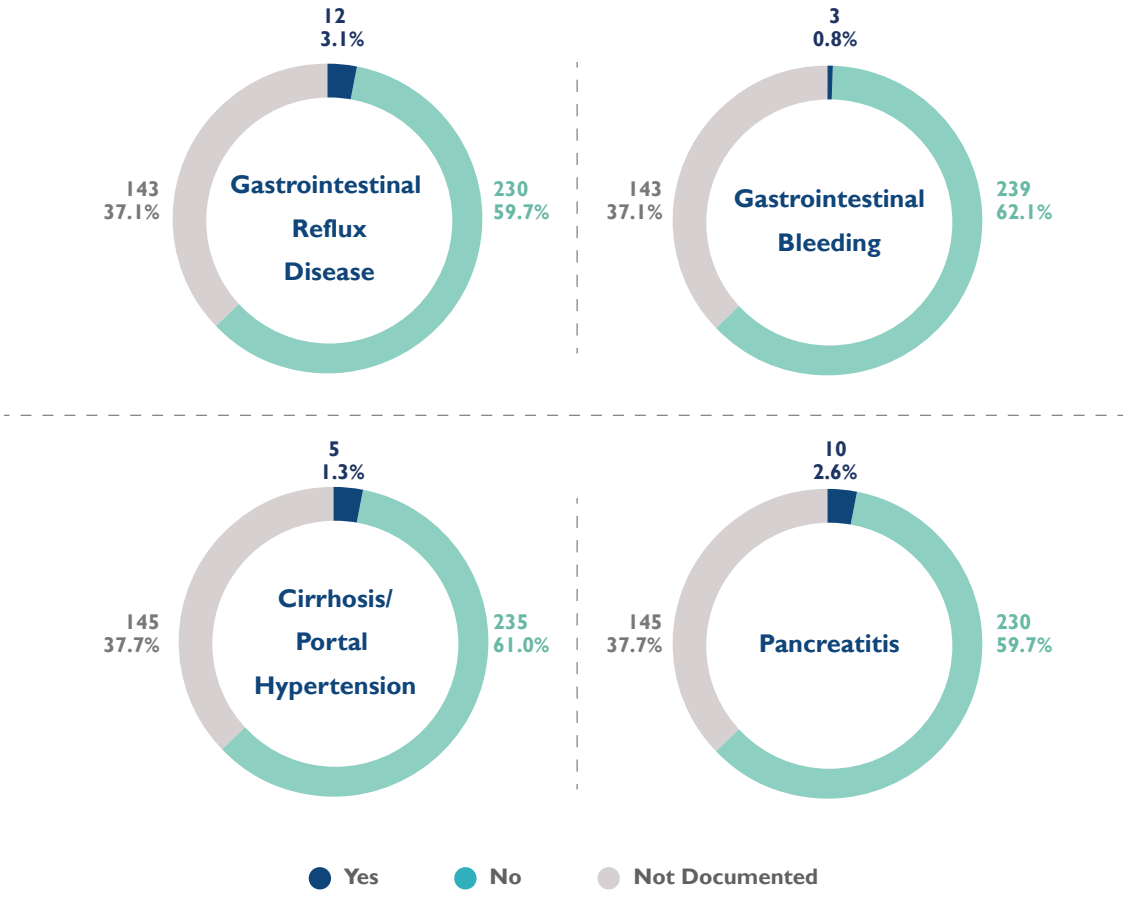
Cystic Fibrosis and Gastrointestinal/Hepatobiliary Manifestations

Assessing the health and functioning of the gastrointestinal (GI) and hepatobiliary systems is of utmost importance in managing CF patients. This includes pancreatic insufficiency, meconium ileus, liver disease, and biliary obstruction, each of which significantly impacts overall health and quality of life. Regular evaluation of GI and hepatobiliary function allows for early detection and intervention, facilitating the optimization of nutritional status, pancreatic enzyme replacement therapy, and appropriate management of liver disease[12].

Distribution of gastrointestinal-related variables

As shown in Table 4 below, approximately 3.1% of CF patients had gastrointestinal reflux disease; 0.8% had gastrointestinal bleeding; 1.3% had cirrhosis/portal hypertension; 2.6% had pancreatitis; and 1.3% had hepatomegaly.

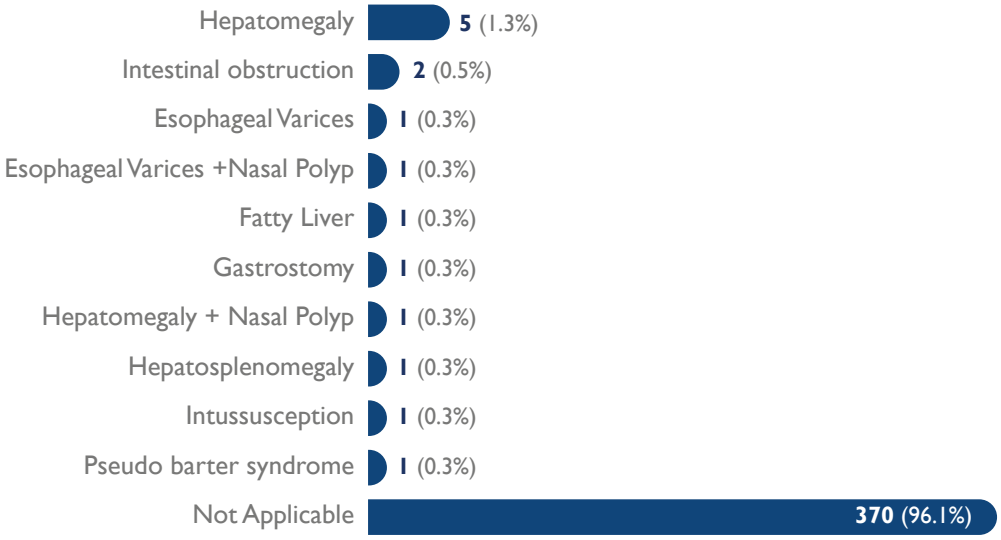
Figure 7: Distribution of Cystic Fibrosis Cases in Jordan According to Gastrointestinal Related Variables



Note: Numbers represent the count of cases associated with cystic fibrosis

12 Stallings, V. A., Stark, L. J., Robinson, K. A., Feranchak, A. P., Quinton, H., & Clinical Practice Guidelines on Growth and Nutrition Subcommittee of the Cystic Fibrosis Foundation, Endocrinology, and Gastroenterology Committees. (2019). Evidence-based practice recommendations for nutrition-related management of children and adults with cystic fibrosis and pancreatic insufficiency: results of a systematic review. Journal of Cystic Fibrosis, 18(6), 803-816.

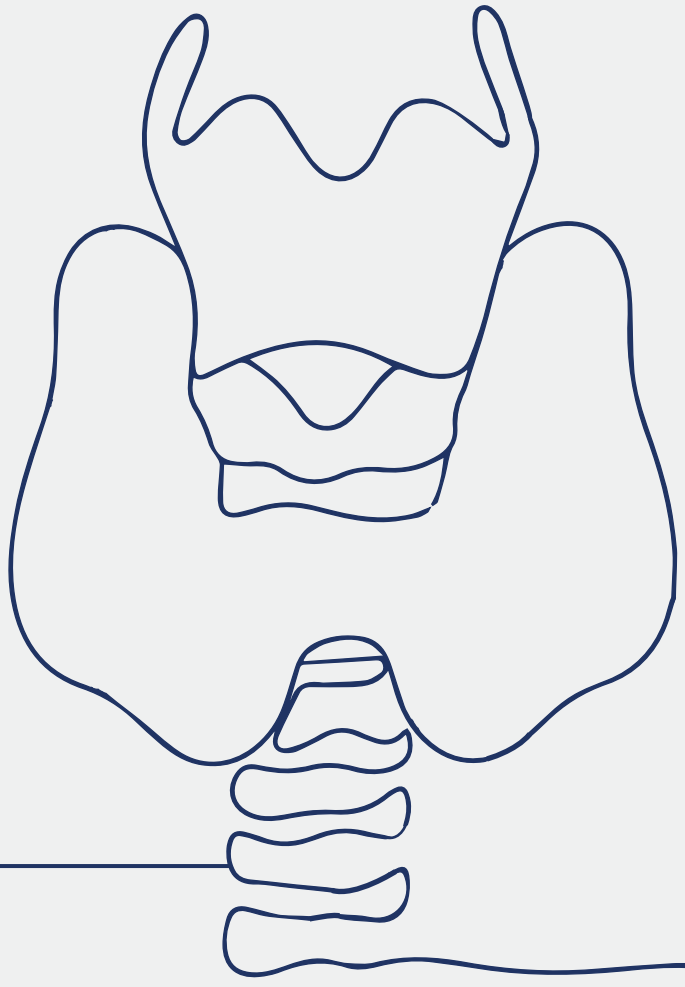
Other Gastrointestinal Diseases



Note: Numbers represent the count of cases associated with cystic fibrosis

5

Cystic Fibrosis Endocrine Manifestations

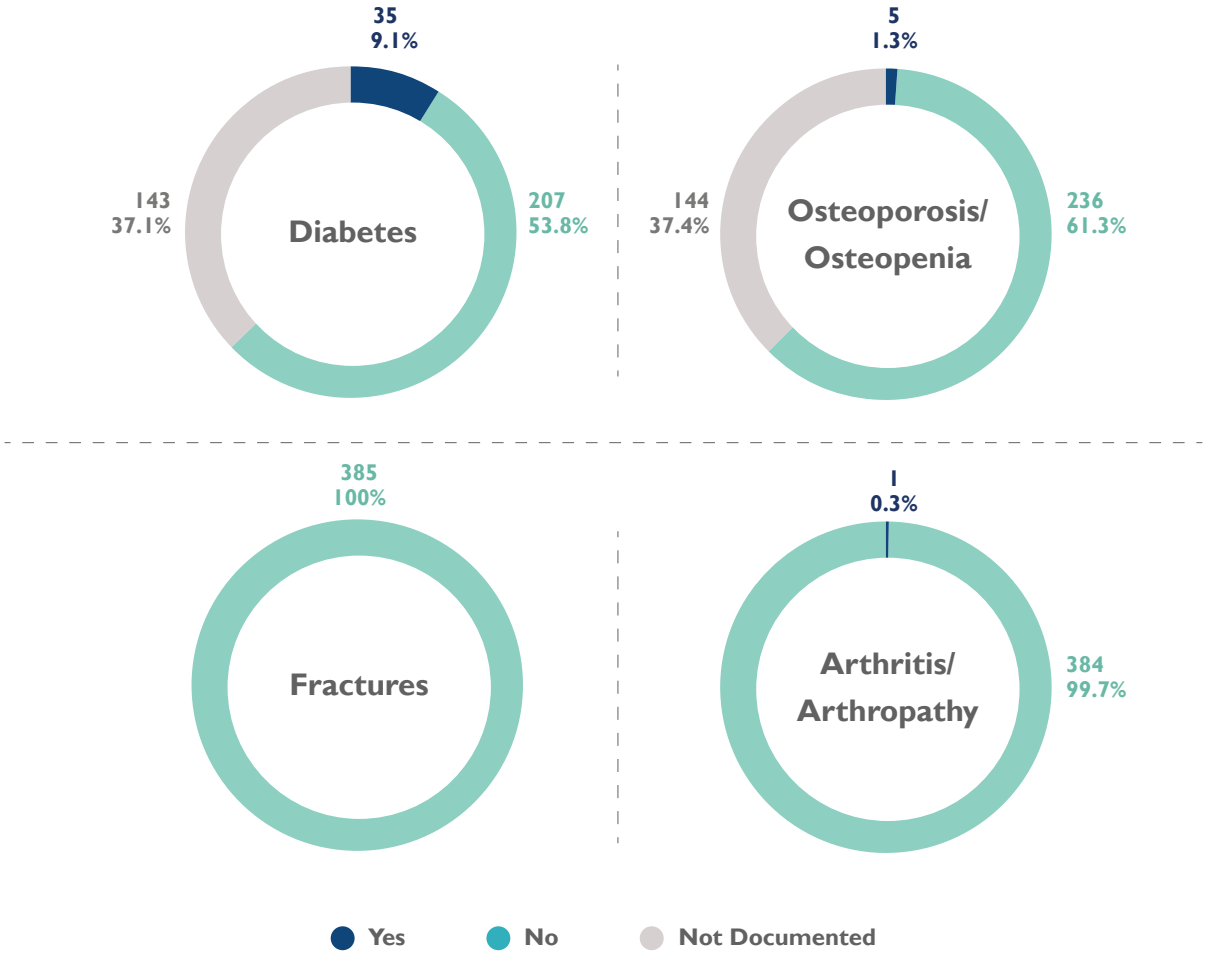


Cystic Fibrosis Endocrine Manifestations

Assessing endocrine functions among CF patients is essential for comprehensive care and management. CF can affect multiple endocrine glands, including the pancreas leading to pancreatic insufficiency, as well as the sweat glands resulting in abnormal salt and water balance. Endocrine complications such as CF-related diabetes (CFRD) and impaired growth can significantly impact the health and quality of life of CF patients. A systematic review published in 2019 in the Journal of Cystic Fibrosis emphasized the importance of regular evaluation and monitoring of endocrine functions in CF. The review highlighted the need for early detection and intervention for conditions like CFRD, growth impairment, and bone health issues to optimize outcomes and provide appropriate management strategies[13]. Furthermore, a study published in 2018 in Pediatric Pulmonology underscored the importance of regular endocrine assessments in CF, particularly for the early identification and management of CFRD[14].

The medical records audit revealed that 9.1% of CF patients had CF-related diabetes, as shown in Table 5. Furthermore, around 2% had orthopedic complications related to diseases such as osteoporosis, osteopenia, and arthritis.

Figure 8: Distribution of Cystic Fibrosis Cases in Jordan According to Endocrine Variables



Note: Numbers represent the count of cases associated with cystic fibrosis

13 Ode, K. L., Frohnert, B., Laguna, T., Phillips, J., Holme, B., Regelmann, W., ... & Moran, A. (2019). Oral glucose tolerance testing in children with cystic fibrosis: An international Delphi survey. Journal of Cystic Fibrosis, 18(6), 786-794.

14 Yen, E. H., Quinton, H., & Borowitz, D. (2018). Better nutritional status in early childhood is associated with improved clinical outcomes and survival in patients with cystic fibrosis. Journal of Pediatrics, 197, 82-88.

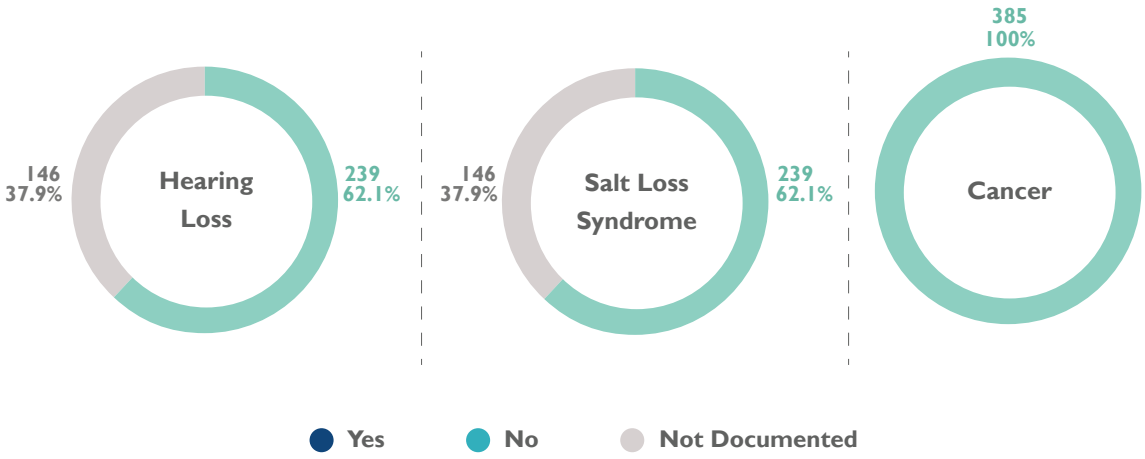
6

Cystic Fibrosis and other Health Variables

Cystic Fibrosis and other Health Variables

Table 6 lists other disease variables found in CF patients in Jordan. No patients had salt loss syndrome, hearing loss, or cancer.

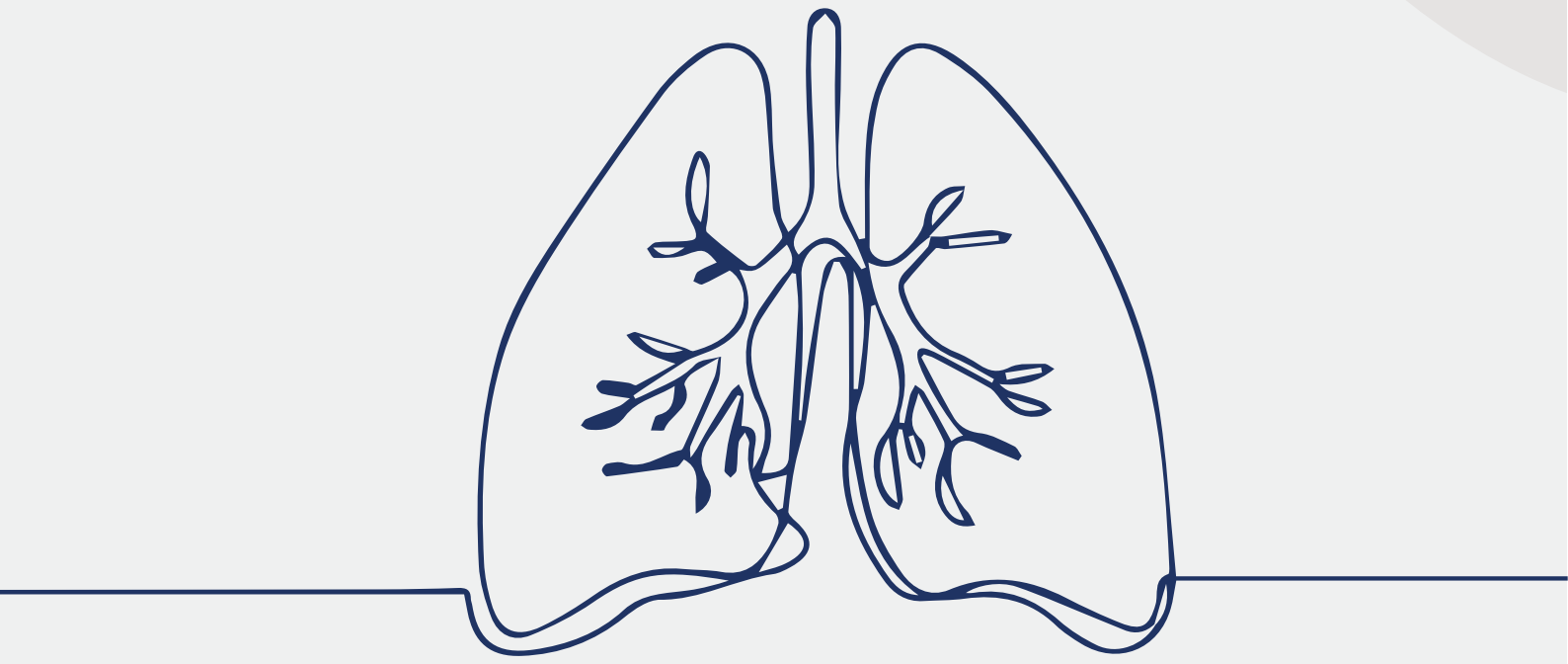
Figure 9: Distribution of Cystic Fibrosis Cases in Jordan According to Other Diseases Variables



Note: Numbers represent the count of cases associated with cystic fibrosis

7

Cystic Fibrosis and Lung Functioning



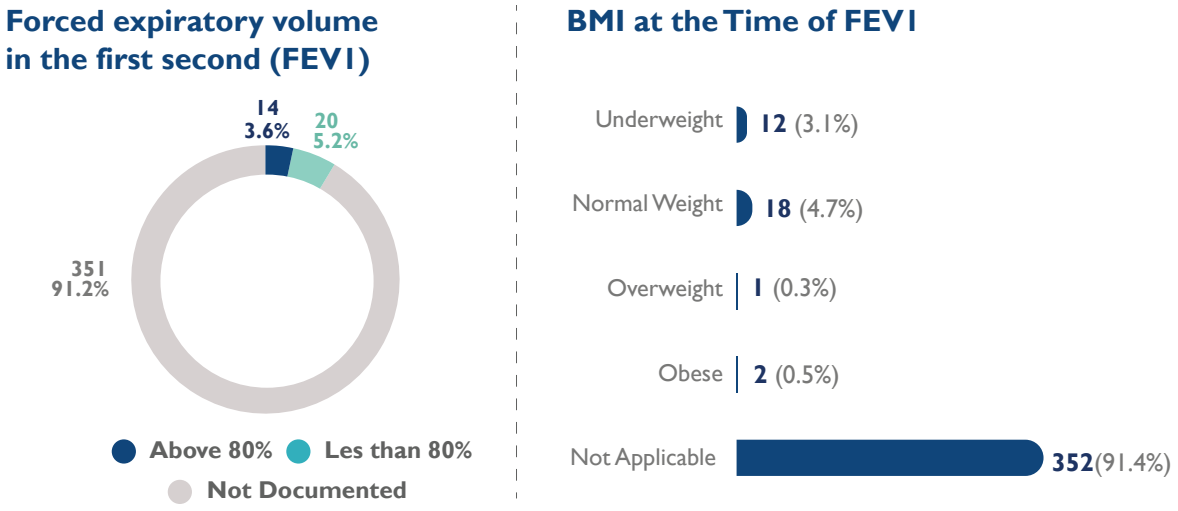
Cystic Fibrosis and Lung Functioning

Pulmonary function tests (PFTs) play a pivotal role in diagnosing and managing CF patients. These tests measure lung capacity and functionality, providing crucial data such as forced expiratory volume in one second (FEV1) and forced vital capacity (FVC). In CF, where thick mucus obstructs the airways leading to respiratory complications, PFTs help in assessing the degree of lung dysfunction, monitoring disease progression, and evaluating the efficacy of treatments. Regular PFT evaluations offer clinicians insights to adjust therapeutic strategies and optimize respiratory health for CF patients.

FEV1 is a vital parameter that reflects the ability of the airways to expel air forcefully within the first second of exhalation. In CF, the presence of airway obstruction and progressive lung damage can lead to decreased FEV1 values. Monitoring FEV1 values over time allows for the early detection of lung function decline, aiding in the diagnosis and management of CF[15].

Table 7 shows that 41% of the documented CF patients FEV1 results had a value above 80%. Unfortunately, 91% of the cases had no documented FEV1 value in the medical record. The BMI at the time of the PFT test shows that patients were either underweight or normal weight.

Figure 10: : Distribution of Cystic Fibrosis Cases in Jordan According to Lung Functioning



Note: Numbers represent the count of cases associated with cystic fibrosis

15 Orenstein, D. M., Spahr, J. E., & Leung, D. H. (2018). Cystic Fibrosis: A Guide for Patient and Family. Springer International Publishing.

8

Distribution of Cystic Fibrosis Cases in Jordan According to Frequency of Seeking Care

Distribution of Cystic Fibrosis Cases in Jordan According to Frequency of Seeking Care

Distribution by number of annual outpatient visits

The average number of annual outpatient visits was three, with patients visiting outpatient clinics up to 18 times annually. Approximately 31% of CF patients visited the outpatient clinic one to five times yearly for follow-up. Around 42% of patients did not visit outpatient clinics at all during 2022.

Distribution by number of hospitalizations

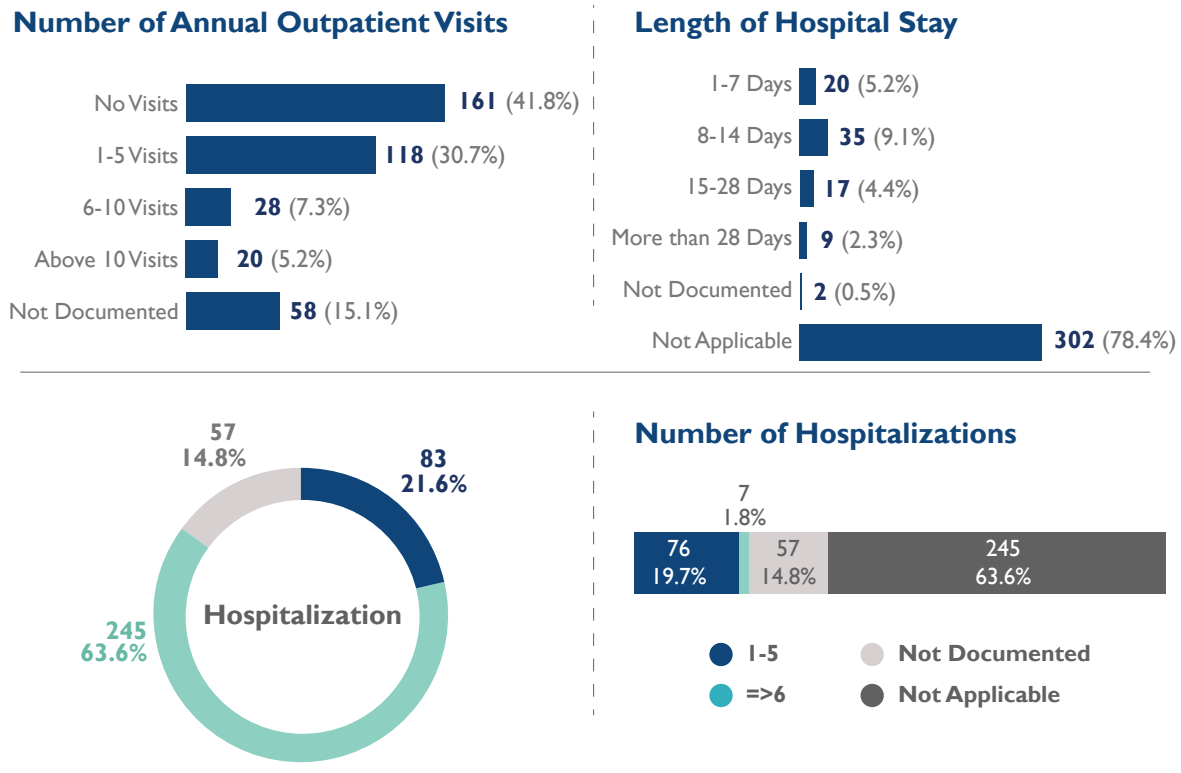
Approximately one-fourth of the patients were admitted to hospitals in 2022. On average, there was one hospitalization per patient during the year, ranging from no hospitalization to a maximum of ten hospitalizations in 2022.

During phone interviews, a significant number of patients or their parents expressed refusal towards hospital admissions. It is crucial to raise awareness about the importance of seeking and receiving timely medical care to enhance the quality of life for these patients.

Distribution by length of hospital stay

The average length of stay for CF patients' admission was approximately 16 to 17 days. Additionally, the majority of patients had a length of stay exceeding eight days. The 25th percentile for the length of stay was eight days, and the median (50th percentile) was 12 days.

Figure 11: Distribution of Cystic Fibrosis Cases in Jordan According to Frequency of Seeking Care



Note: Numbers represent the count of cases associated with cystic fibrosis

9

Pulmonary Treatment of Cystic Fibrosis

Pulmonary Treatment of Cystic Fibrosis

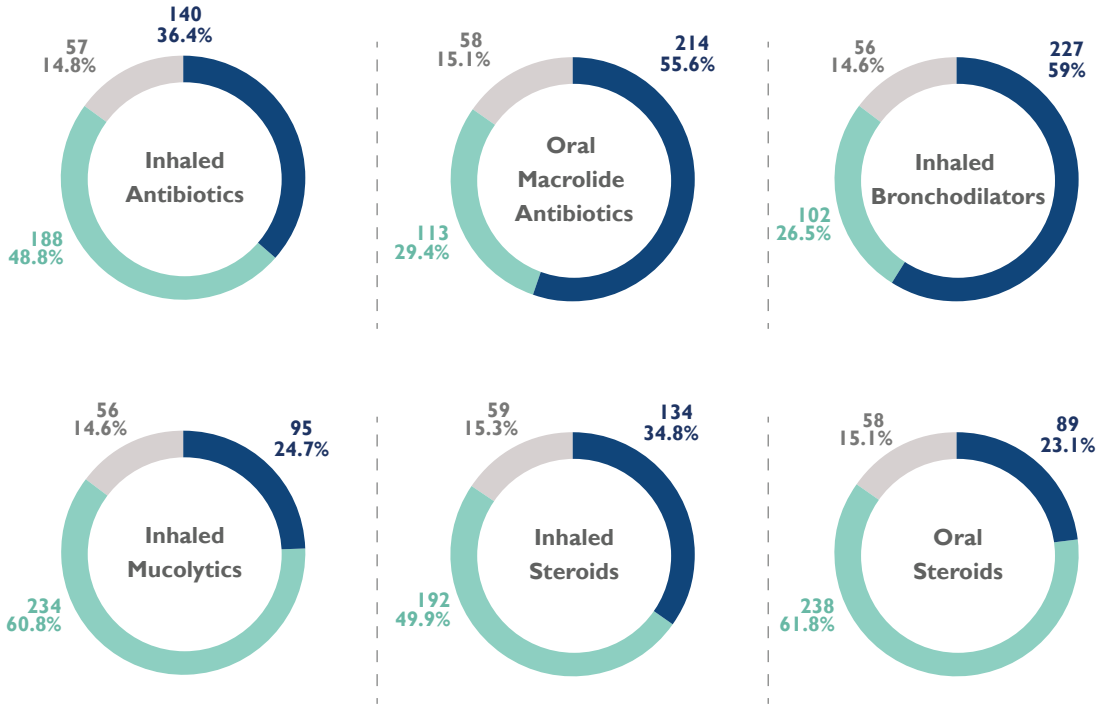
Around 36% of CF cases received inhaled antibiotics which play a crucial role in managing CF patients, particularly those with chronic lung infections. By directly targeting the bacteria in the lungs, inhaled antibiotics help to reduce bacterial load, control infection, and slow down the progression of lung damage. Tobramycin and aztreonam are commonly used inhaled antibiotics among CF patients in Jordan with pseudomonas aeruginosa infections, the most commonly reported bacterial infection[16].

Out of 327 documented cases of bacterial infection, 214 patients (65%) received oral macrolide antibiotic. Around 60% of patients received inhaled bronchodilator, 25% received inhaled mucolytics like hypertonic saline, 35% received inhaled steroids, and just six patients received non-invasive ventilation such as CPAP/BiPAP at home.

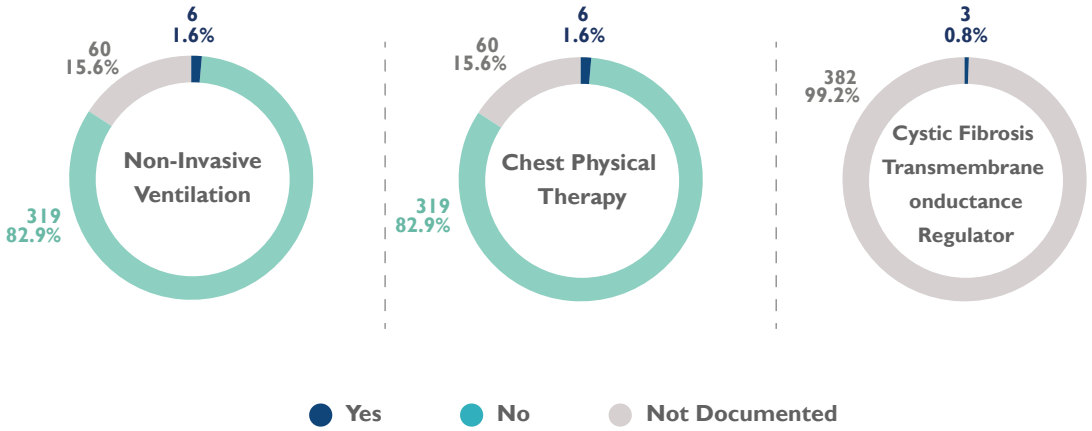
Six patients regularly received chest physiotherapy (CPT) from healthcare providers or family caregivers. This raises the need for increasing the awareness of these patients/families about the importance of CPT.

Three CF patients received CFTR modulators, which are not available in Jordan. These medications target the defective CFTR protein, which is responsible for the underlying cause of CF. CFTR modulators can improve lung function and overall health in patients with specific CF mutations.

Figure 12: Distribution of Cystic Fibrosis Cases in Jordan According to Pulmonary Treatment



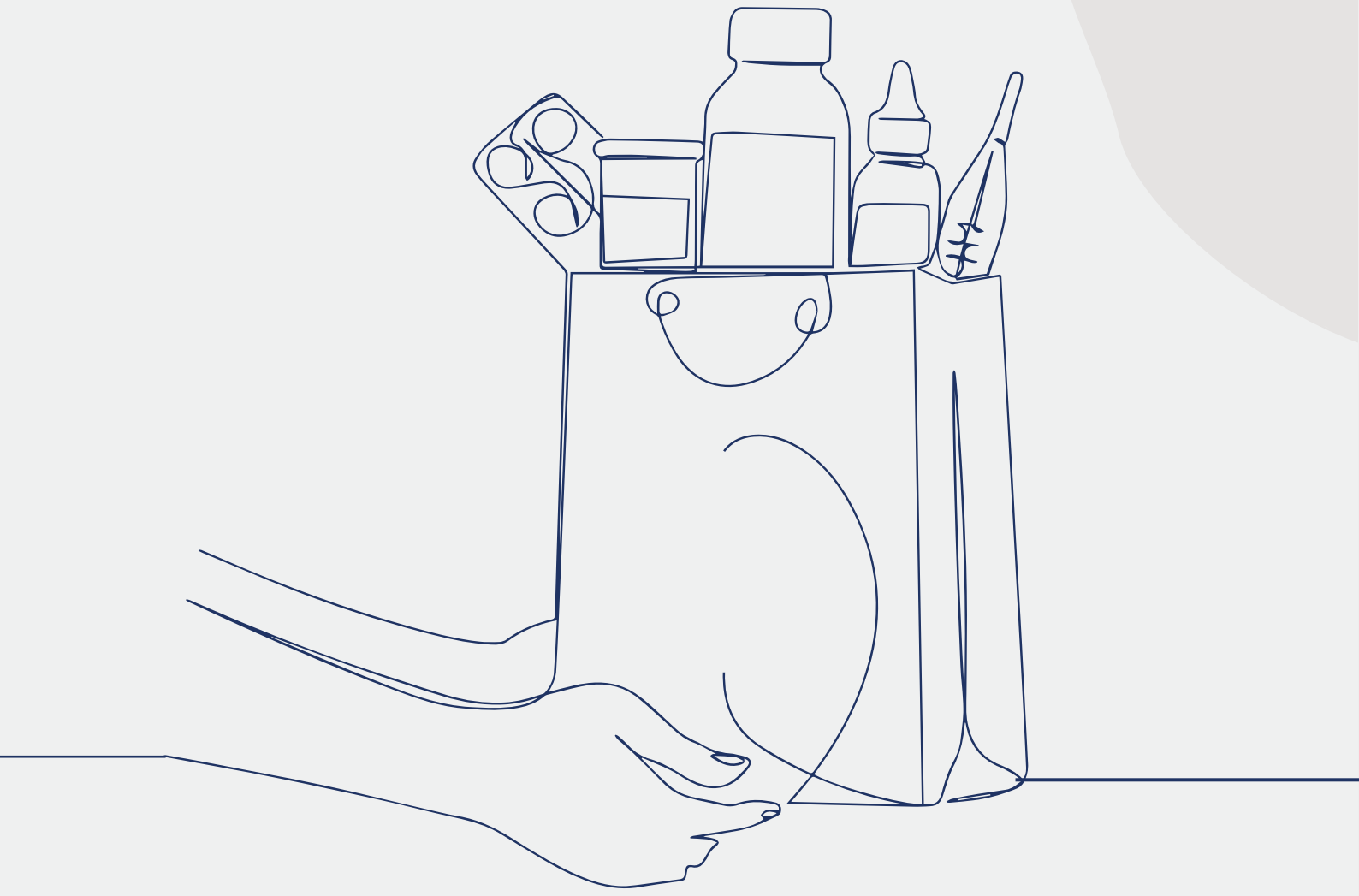
16 Rowe, S. M., et al. New England Journal of Medicine, 373(3), 220-231, 2015; Donaldson, S. H., et al. New England Journal of Medicine, 372(21), 1985-1993, 2015; Ramsey, B. W., et al. New England Journal of Medicine, 373(3), 232-242, 2015



Note: Numbers represent the count of cases associated with cystic fibrosis

10

Gastrointestinal Treatment of Cystic Fibrosis

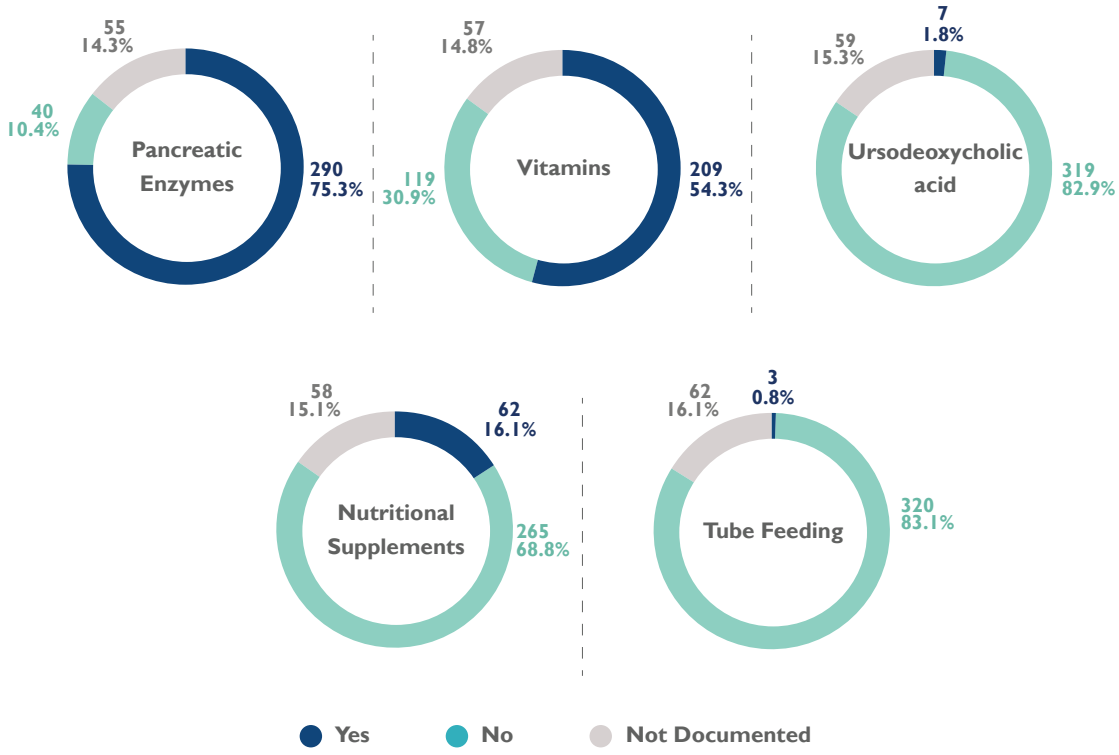


Gastrointestinal Treatment of Cystic Fibrosis

The gastrointestinal treatment of CF patients is of paramount importance in managing the disease and promoting optimal nutrition and growth. CF can lead to various GI complications, including pancreatic insufficiency and malabsorption of nutrients. Enzyme replacement therapy is a cornerstone in treating exocrine pancreatic insufficiency, with pancreatic enzyme supplements being prescribed to aid in the digestion and absorption of essential nutrients. Additionally, some patients may develop CF-related liver disease, necessitating specialized management and monitoring. Nutritional support and personalized dietary plans are crucial in maintaining adequate weight and growth in CF patients. A multidisciplinary approach involving gastroenterologists, dieticians, and CF specialists is essential in addressing the diverse GI challenges faced by individuals with CF .

As shown in Table 10, the majority of patients received pancreatic enzyme medications. About half of the patients received multivitamins, mainly vitamins A, B, C, D, E, and K, and 16% received nutritional supplements. Just three patients, who were infants, received tube feeding.

Figure 13: Distribution of Cystic Fibrosis Cases in Jordan According to Gastrointestinal Treatment



Note: Numbers represent the count of cases associated with cystic fibrosis

Data Limitations

Collecting data for this report presented several challenges and limitations due to various factors, including:

- Lack of a national CF registry: Establishing a comprehensive and accurate live CF registry from scratch requires significant effort and resources. Setting up data collection mechanisms, obtaining consent from patients and healthcare providers, and ensuring data quality can be time-consuming and complex.
- Incomplete historical data: Gathering historical data for CF patients was challenging, especially in cases where the medical records were not well maintained and patients sought treatment from multiple healthcare providers among different health sectors in Jordan. This resulted in gaps in the patient's medical history, potentially affecting the accuracy of the report's findings.
- Data validation and accuracy: Ensuring the accuracy and validity of collected data is crucial. Inconsistencies in recording practices could lead to inaccuracies in the report. Establishing a live electronic registry will overcome this limitation.
- Data standardization: CF data were collected from various sources, including clinics, hospitals, and CF centers. Each source used different data formats or terminologies, making data standardization challenging. Harmonizing data across different sources is essential for meaningful analysis and comparison. It is recommended to have a standardized template for filling in CF patients' information, whether using paper or electronic medical records. Again, a live electronic CF registry platform will help to ensure standardized CF data for Jordan.
- Patient confidentiality and consent: Collecting patient data for a CF report requires adherence to strict privacy regulations and obtaining informed consent from participants. Maintaining patient confidentiality while aggregating data for analysis can be a complex process. Reaching all patients through phone calls may not be feasible. There is a need to obtain consent through the clinics that the patients visit regularly.
- Bias in data collection: Bias in data collection occurred for certain patient groups who are overrepresented or underrepresented in the registry. For example, patients with severe or advanced CF may be more likely to seek medical attention, leading to an uneven representation in the data.

Addressing these challenges and limitations requires a well-organized data collection process, a collaboration between healthcare providers across all health sectors in Jordan, and a commitment to data quality and accuracy. It is essential to continuously refine data collection methods to improve the reliability and usefulness of the CF report for guiding treatment decisions and advancing CF research for Jordan.

Recommendations

1. Standardized data collection: Implement standardized data collection protocols across all healthcare facilities and CF centers participating in the registry by using the electronic registry platforms to fill the data by the healthcare workers. This ensures consistency and comparability of data, making it easier to analyze trends and draw meaningful conclusions.
2. Comprehensive patient information: Expand the scope of data collection to include a comprehensive set of patient information, such as demographics, clinical characteristics, genetic mutations, lung function measures in detail, comorbidities, treatments, and outcomes. This will provide a holistic view of the CF population in Jordan and support diverse research initiatives. Clerks and admission staff need to be trained and required to record and update patient data.
3. Real-time data updates: Encourage regular and timely data updates from participating healthcare facilities to maintain accurate and up-to-date information by using the live electronic national CF registry platform to update patients' data regularly. Real-time data collection can offer a more accurate reflection of the current CF landscape and support swift decision-making.
4. Center of excellence for CF patients: Establishing this center will ensure comprehensive care for CF patients in collaboration with the MOH satellite CF centers.
5. Longitudinal data analysis: Emphasize the importance of longitudinal data analysis by tracking the progression of CF patients over time in Jordan. This will enable healthcare providers and researchers to understand disease trajectories, treatment responses, and long-term outcomes. It could help the decision-makers at Jordan's MOH to use such data in their decisions.
6. Data validation and quality assurance: Implement rigorous data validation and quality assurance procedures to ensure the reliability and integrity of data. Regular audits and checks should be conducted to identify and correct any inaccuracies or inconsistencies in the electronic registry platform. A maintenance contract with an information system vendor is needed.
7. Data security and privacy: Prioritize data security and patient privacy by adopting robust encryption and access controls. Ensure compliance with relevant data protection regulations and maintain patient confidentiality at all times.
8. Collaboration and research initiatives: Encourage collaboration among researchers and institutions to leverage Jordan's CF registry data for research initiatives. Facilitate data-sharing agreements while safeguarding patient privacy, fostering a collaborative approach to advancing CF knowledge and treatment. For example, connect the national registry with the Middle East and/or International Cystic Fibrosis Associations. Patient engagement: Involve CF patients and their families in the data collection and reporting process. Seek their feedback to ensure the registry captures relevant patient-centered outcomes and reflects the real-life challenges faced by the CF community, like the Jordan Cystic Fibrosis Friends Charity.
9. Comparative analysis: Perform comparative analysis with regional and international CF registries to gain insights into global CF trends, treatment strategies, and outcomes. This can help identify best practices and areas for improvement in CF management.
10. User-friendly reporting: Design the annual report in a user-friendly format with clear visualizations and concise summaries. The report should cater to diverse audiences, including healthcare professionals, researchers, policymakers, and patient advocacy groups.
11. Transparency and accessibility: Make the annual report publicly accessible to promote transparency and accountability. Consider sharing the report on the organization's website and other relevant platforms to reach a wider audience.

Contributors

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Appendices

Appendix A: Cystic Fibrosis Patient Consent Form

نموذج موافقة المريض أو قريب المريض لغايات إنشاء السجل الوطني لمرض التليف الكيسي واستخدام معلومات السجل لغايات تحسين الخدمات الصحية المقدمة لمرضى التليف الكيسي

هدف الموافقة: تهدف هذه الموافقة إلى مساهمتك بإنشاء سجل وطني لمرض التليف الكيسي في الأردن والذي يتضمن دراسة الوضع الحالي للمرضى المشخصين بهذا المرض والتحديات التي تواجههم في التشخيص ورحلة العلاج. بالإضافة إلى إيجاد سبل الارتقاء بجودة الخدمات الصحية المقدمة لهم عن طريق بيانات السجل.

لماذا تم اختياري/اختيار المريض للمشاركة في إنشاء السجل الوطني لمرضى التليف الكيسي؟ لقد تم اختيارك/اختيار المريض لأنك من الأشخاص الذين تنطبق عليهم الشروط التالية، وهي أنك مريض أو وصي عن مريض تم تشخيصه بمرض التليف الكيسي، إذا كنت وصي عن المريض وعمرك ١٨ سنة أو أكثر أوعمر المريض أقل من ١٨ سنة، ولديك القدرة على فهم اللغة العربية.

ماذا علي أن افعل؟ عند موافقتك المشاركة يطلب منك الاستجابة لبعض المعلومات الديموغرافية من قبل جامعي البيانات المخولين بجمع هذه المعلومات بالتعاون مع وزارة الصحة، وسيقوم جامعي البيانات بمساعدتك في حال وجود أي صعوبة أثناء الإجابة.

هل ستعامل المعلومات التي يتم جمعها بسرية؟ كل المعلومات المقدمة لغرض إنشاء السجل الوطني لمرض التليف الكيسي ستعامل بسرية تامة، من خلال عدم كتابة اسمك/ اسم المريض وعدم مشاركة أي معلومات دالة عن المريض. ولن يطلع على محتوى المعلومات إلا المخولين من وزارة الصحة عن طريق رمز أو رقم يمثل المريض بدون ذكر اسمه أو أي معلومات عنه.

هل يترتب أي مسؤولية من مشاركتي في إنشاء السجل؟ لا يوجد أي مسؤولية أو تداعيات مترتبة على مشاركتك في إنشاء السجل الوطني لمرض التليف الكيسي.

هل لي الحرية الشخصية بالانسحاب من المشاركة؟ يمكنك الانسحاب من المشاركة في أي وقت بدون أية عواقب ولن يؤثر ذلك بأي شكل على الرعاية الصحية المقدمة للمريض في كافة القطاعات الصحية في الأردن.

كم أحتاج من الوقت لتعبئة المعلومات؟ إذا قررت الاشتراك فإن مدة تعبئة المعلومات المطلوبة هاتفيا تتراوح من ٥-١٠ دقائق.

هل يمكنني الحصول على التقرير الإحصائي الوطني بعد الانتهاء من إنشاء سجل مرض التليف الكيسي؟ إذا أردت أي استفسار عن حيثيات السجل في أي وقت يمكنك التواصل مع وزارة الصحة/ مديرية الأمراض غير السارية.

تصريح بالموافقة: لقد تم توضيح كل النقاط السابقة بشكل مفصل. وفهمت أن لي الحق بطرح أي سؤال في أي وقت كان. وأن هذه الأسئلة ستجاب من خلال جامعي البيانات المخولين من وزارة الصحة.

بموافقتي الشفهية هاتفيا على هذا النموذج، تعني موافقتي على أن أكون كمريض او كوصي عنه أحد المشاركين في إنشاء السجل الوطني لمرض التليف الكيسي في الأردن.

اسم المريض: **رقم الهاتف:**

اسم وصي المريض ودرجة القرابة:

اسم جامع البيانات:

أسماء المستشفيات التي يتعالج بها المريض:

التاريخ :

Appendix B Data Collection Tool

JORDAN CYSTIC FIBROSIS REGISTRY DATA TOOL

Source of data	Pediatrician	Parents	Patient	
Hospital name				
Health Insurance	Yes (Type: ……..)		No	
1- Patient's Personal and Demographic Data				
Demographics and Patient's Personal Data				
Nationality	Jordanian		Non-Jordanian (Specify: ……..……..….)	
Patient National ID/ Personal ID				
Date of birth (MM/DD/YYYY)				
Full Name				
Gender	Male		Female	
Marital Status	Single	Married	Divorced/widow	
Education Level	Illiterate	Basic	Secondary	Higher than secondary
Address				
2- Diagnosis				
Patient was diagnosed by Select all that apply?	1. Prenatal Diagnosis	2.Newborn Screening	3.Clinical manifestations Respiratory GI manifestations / Pancreatic Insufficiency	4.Meconium Ileus at birth
	5.Family History or positive sibling		7. other Specify (……..…)	
Age at Diagnosis Age in month if <2yrs Age in yrs if >2yrs				
Sweat test type?	Chloride Titration	Conductivity	Unknown	
Diagnostic sweat chloride value (mmol/L)?				
Fecal Elastase value, (microg/g of stool)?				
Was the patient genotyped?	Mutation 1	Mutation 2	Not genotyped	
Genotyping date: (DD MM YYYY)				

Source of data	Pediatrician	Parents	Patient
3- Health and Functioning			
3.1 Respiratory Infections			
Number of sputum and BAL/bronchoscopy cultures per year			
Microbiology: Positive Cultures Select all that apply:	**Fungi** • Candida • Aspergillus • Others • (specify ……..…)	**Bacteria:** • Pseudomonas: mucoid and non-Mucoid • Staphylococcus aureus MSSA • Haemophilus influenza • Burkholderia cepacia • Stenotrophomonas meltophilia • Escherichia coli • Staphylococcus aureus MRSA • Serratia marcescens • Klebsiella • Non-tuberculous mycobacterium • Other (specify……)	**Unknown**
3.2 Other Medical Complications			
Pulmonary/Airways Select all that apply?	• Major hemoptysis • Bronchiectasis • Pneumothorax • Fibrosis • Respiratory failure • Others (specify ……..…..)		
Gastrointestinal/ Hepatobiliary Select all that apply?	• GERD Gastroesophageal Reflux Disease • Gastrointestinal bleeding • Abnormal Liver Function test • Cirrhosis or portal Hypertension • Pancreatitis • Others (specify……)		
Endocrine	• Diabetes • Osteoporosis/Osteopenia • Fractures • Arthritis/Arthropathy • Others (specify……)		
Others	• Hearing loss • Salt loss syndrome • Cancer • other complications (specify……..…)		

3.3 Lung Function (Anthropometric and Spirometry Measurements)			
The best-recorded FEV1 in the year 2022			
FEV1 date			
Obstructive pattern	Mild	Moderate	Severe
Mixed pattern	Mild	Moderate	Severe
3.4 Height and Weight (at the time of FEV1 best value)			
Height			
Weight			
4. Treatment of Cystic Fibrosis			
4.1 number of clinic visits per this year of data collection			
4.1 Pulmonary Therapy			
Inhaled pulmonary antibiotics? Gentamycin, Tobramycin, Amikacin. Colistin	Yes	No	Unknown
Inhaled bronchodilators?	Yes	No	Unknown
Inhaled mucolytic therapies?	Yes	No	Unknown
Select all mucolytics that apply:	Dornase Alfa or Pulmozyme, rhodanese	Hypertonic saline	N-Acetylcysteine
Oral macrolide antibiotics?	Yes	No	Unknown
4.2 Other Therapies			
Oral steroids	Yes	No	Unknown
Inhaled steroids?	Yes	No	Unknown
Non-invasive ventilation (i.e., assisted breathing, BiPap, CPAP, etc)?	Yes	No	Unknown
Oxygen therapy?	Yes (specify ……)	No	Unknown
Chest Physiotherapy?	Yes (specify ……)	No	
Cystic Fibrosis Transmembrane Conductance Regulator Modulators?	Yes	No	Unknown
Specify the last CFTR modulator used?	Single therapy, e.g., ivacaftor)	Double therapy, e.g. teza/iva or lum/iva)	Triple therapy, e.g., elexa/teza/iva)
Was post CFTR modulator sweat value measured?	Yes	No	Unknown

Sweat test type (after CFTR modulator)?	Conductivity		Chloride Titration
Post CFTR modulator sweat chloride value (mmol/L)?			
4.3 GI Medications			
Use of pancreatic enzymes? (Creon)	Yes	No	Unknown
Cystic Fibrosis specific vitamins?	Yes	No	Unknown
Ursodeoxycholic Acid?	Yes	No	Unknown
Nutritional Supplements Pediasure milk, or ensure for adults	Yes, through tube feeding	yes, through an oral supplement	Unknown
Tube feedings	Naso/Orogastric tube	Gastrostomy tube	Total Parenteral Nutrition
4.5 Diabetic Therapies			
Was the patient diagnosed with Cystic Fibrosis Related Diabetes?	Yes	No	Unknown
What diabetic therapies were used?	Insulin	Oral hypoglycemic agent	None
4.6 Hospital Treatment			
Number of hospitalizations related to CF per this year			
Average Length of Hospital Stay per this year.			
4.7 Non-Transplant Surgery			

- IV access devices
- Gall bladder disease
- Gastrostomy
- Intestinal obstruction
- Nasal (polyp)
- Others (specify……..)

5. Transplantation?		
Has the patient ever had transplantation?	Yes (liver or Lung)	No

Appendix C: Summary Statistics of Numeric Variables

Variable	N	Mean	Min	Max
Age in Years	355	17.23	0.65	52.19
Age at Diagnosis in Years	212	2.57	0	33
Sweat Cl value (mmol/L)	222	95.28	45	195
N Cx/yr	168	2.23	0	12
FEV 1	34	71.24	34	115
N Visits-yr	327	2.47	0	18
No of hospitalizations	328	0.55	0	10
Avg Length of stay	81	16.32	3	90
Body Mass Index Values	33	20.24	11.83	39.41

Appendix D: List of Mutations found in CF patients in Jordan

1. W1282X homozygous
2. N1303 homozygous
3. MR 334W homozygous
4. MR 334W heterozygous
5. MM1303k Heterozygous
6. MG542X homozygous
7. M2183 A.G homozygous
8. M.G85E homozygous
9. M.G85E heterozygous
10. .M 2789+56A homozygous
11. G84 homozygous
12. Delta F 508
13. Delta 2184
14. Delta 1677
15. Delta 1603+2875
16. 1525-1 G>A homozygous
17. 117 heterozygous

Appendix E: List of Tables on Cystic Fibrosis Cases in Jordan

Table 1: Distribution of Cystic Fibrosis Cases in Jordan According to Background Variables

Background Variable	Number of Cases	Percent
Governorate		
Ajloun	15	3.9
Amman	136	35.3
Aqaba	3	0.8
Balqa	12	3.1
Irbid	100	26.0
Jerash	5	1.3
Karak	16	4.2
Madaba	2	0.5
Mafraq	10	2.6
Tafilah	2	0.5
Zarqa	33	8.6
Not Documented	51	13.3
Current Age Groups		
<10 years	62	16.1
>10 and <20	183	47.5
>20 and <30 years	86	22.3
30 years and above	24	6.2
Not Documented	30	7.8
Sex		
Female	167	43.4
Male	218	56.6
Marital Status		
Single	381	99.0
Married	4	1.0
Educational Level		
Illiterate	24	6.2
Basic	97	25.2
Secondary	85	22.1
Higher than Secondary	21	5.5
Not Documented	158	41.0
Nationality		
Jordanian	375	97.4
Syrian	6	1.6
Other	4	1.0
Health Insurance		
Yes	377	97.9
No	8	2.1
Total	385	100

Table 2: Distribution of Cystic Fibrosis Cases in Jordan According to Diagnosis Related Variables

Variable	Number of Cases	Percent
Age at Diagnosis		
At Birth	16	4.2
Less than 1 Year	96	24.9
1-9 Years	90	23.4
10-20 Years	8	2.1
More than 20 Years	2	0.5
Not Documented	173	44.9
Prenatal Diagnosis		
No	385	100.0
Newborn Screening		
No	385	100.0
Clinical Manifestations		
Yes	348	90.4
No	3	0.8
Not Documented	34	8.8
Meconium Ileus		
Yes	19	4.9
No	366	95.1
Sweating Test		
Chloride Titration	182	47.3
Conductivity	70	18.2
Not Documented	133	34.6
Chloride Value (Sweat Test)		
30-59 mmol/L	21	5.5
=>60 mmol/L	201	52.2
Not Documented	163	42.3
Genotyping		
Yes	114	29.6
No	167	43.4
Not Documented	104	27.0
Mutation Testing		
Positive	56	14.6
Negative	2	0.5
Documented	327	84.9
Total	385	100

Table 3: Distribution of Cystic Fibrosis Cases in Jordan According to Respiratory Variables

Variable	Number of Cases	Percent
Annual Chest X-Ray		
None	17	4.4
1-3 times	110	28.6
4-6 times	37	9.6
More than 6 times	4	1.0
Not Documented	217	56.4
Microbiology Testing		
Bacteria	182	47.3
Bacteria and Fungi	3	0.8
Fungi	5	1.3
No Growth	4	1.0
Not Documented	191	49.6
Hemoptysis		
Yes	97	25.19
No	147	38.18
Not Documented	141	36.62
Lung Fibrosis		
Yes	7	1.8
No	237	61.6
Not Documented	141	36.6
Respiratory Failure		
Yes	4	1.0
No	239	62.1
Not Documented	142	36.9
Other Respiratory Diseases		
Nasal polyps	17	4.4
Pansinusitis	1	0.3
Pleural effusion	1	0.3
Pulmonary Hypertension	1	0.3
Not Applicable	365	94.8
Total	385	100

Table 4: Distribution of Cystic Fibrosis Cases in Jordan According to Gastrointestinal Related Variables

Variable	Number of Cases	Percent
Gastrointestinal Reflux Disease		
Yes	12	3.1
No	230	59.7
Not Documented	143	37.1
Gastrointestinal Bleeding		
Yes	3	0.8
No	239	62.1
Not Documented	143	37.1
Cirrhosis/Portal Hypertension		
Yes	5	1.3
No	235	61.0
Not Documented	145	37.7
Pancreatitis		
Yes	10	2.6
No	230	59.7
Not Documented	145	37.7
Other Gastrointestinal Diseases		
Hepatomegaly	5	1.3
Intestinal obstruction	2	0.5
Esophageal Varices	1	0.3
Esophageal Varices + Nasal Polyp	1	0.3
Fatty Liver	1	0.3
Gastrostomy	1	0.3
Hepatomegaly + Nasal Polyp	1	0.3
Hepatosplenomegaly	1	0.3
Intussusception	1	0.3
Pseudo barter syndrome	1	0.3
Not Applicable	370	96.1
Total	385	100

Table 5: Distribution of Cystic Fibrosis Cases in Jordan According to Endocrine Variables

Variable	Number of Cases	Percent
Diabetes		
Yes	35	9.1
No	207	53.8
Not Documented	143	37.1
Osteoporosis/Osteopenia		
Yes	5	1.3
No	236	61.3
Not Documented	144	37.4
Fractures		
No	385	100
Arthritis/Arthropathy		
Yes	1	0.3
No	384	99.7
Total	385	100

Table 6: Distribution of Cystic Fibrosis Cases in Jordan According to Other Diseases Variables

Variable	**Number of Cases**	**Percent**
Hearing Loss		
No	239	62.1
Not Documented	146	37.9
Salt Loss Syndrome		
No	239	62.1
Not Documented	146	37.9
Cancer		
No	385	100
Total	385	100

Table 7: Distribution of Cystic Fibrosis Cases in Jordan According to Lung Functioning

Variable	Number of Cases	Percent
Forced expiratory volume in the first second (FEV1)		
Above 80%	14	3.6
Less than 80%	20	5.2
Not Documented	351	91.2
BMI at the Time of FEV1		
Underweight	12	3.1
Normal Weight	18	4.7
Overweight	1	0.3
Obese	2	0.5
Not Applicable	352	91.4
Total	385	100

Table 8: Distribution of Cystic Fibrosis Cases in Jordan According to Frequency of Seeking Care

Variable	Number of Cases	Percent
Number of Annual Outpatient Visits		
No Visits	161	41.8
1-5 Visits	118	30.7
6-10 Visits	28	7.3
Above 10 Visits	20	5.2
Not Documented	58	15.1
Hospitalization		
Yes	83	21.6
No	245	63.6
Not Documented	57	14.8
Number of Hospitalizations		
1-5	76	19.7
=>6	7	1.8
Not Documented	57	14.8
Not Applicable	245	63.6
Length of Hospital Stay		
1-7 Days	20	5.2
8-14 Days	35	9.1
15-28 Days	17	4.4
More than 28 Days	9	2.3
Not Documented	2	0.5
Not Applicable	302	78.4
Total	385	100

Table 9: Distribution of Cystic Fibrosis Cases in Jordan According to Pulmonary Treatment

Variable	Number of Cases	Percent
Inhaled Antibiotics		
Yes	140	36.4
No	188	48.8
Not Documented	57	14.8
Oral Macrolide Antibiotics		
Yes	214	55.6
No	113	29.4
Not Documented	58	15.1
Inhaled Bronchodilators		
Yes	227	59.0
No	102	26.5
Not Documented	56	14.6
Inhaled Mucolytics		
Yes	95	24.7
No	234	60.8
Not Documented	56	14.6
Inhaled Steroids		
Yes	134	34.8
No	192	49.9
Not Documented	59	15.3
Oral Steroids		
Yes	89	23.1
No	238	61.8
Not Documented	58	15.1
Non-Invasive Ventilation		
Yes	6	1.6
No	319	82.9
Not Documented	60	15.6
Chest Physical Therapy		
Yes	6	1.6
No	319	82.9
Not Documented	60	15.6
Cystic Fibrosis Transmembrane Conductance Regulator		
Yes	3	0.8
No	382	99.2
Total	385	100

Table 10: Distribution of Cystic Fibrosis Cases in Jordan According to Gastrointestinal Treatment

Variable	Number of Cases	Percent
Pancreatic Enzymes		
Yes	290	75.3
No	40	10.4
Not Documented	55	14.3
Vitamins		
Yes	209	54.3
No	119	30.9
Not Documented	57	14.8
Ursodeoxycholic acid		
Yes	7	1.8
No	319	82.9
Not Documented	59	15.3
Nutritional Supplements		
Yes	62	16.1
No	265	68.8
Not Documented	58	15.1
Tube Feeding		
Yes	3	0.8
No	320	83.1
Not Documented	62	16.1
Total	385	100

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