Navigating Adolescence with Cystic Fibrosis: A systematic review on impact of cystic fibrosis with teenagers.

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Abstract

Cystic fibrosis is a challenging disease which creates many complications. The impact caused from cystic fibrosis on an adolescent is a challenging phase for their lives. Negative impacts like the society's pressure, anxiety and depression are few common reasons that are being unspoken and ignored due to complexity of the disease. This study is aimed to identify the necessary gaps and educate the necessary policy makers and CF center health care team to have a better understanding about the context of the aftermath of cystic fibrosis and enhance better patient care. More over this systematic review also points out the main trends in this field of study. Qualitative study and quantitative are prominently used research methods to gain an understanding the about the gaps in the research. However, there is still a scarcity of quantitative and mixed research methods. Furthermore, limitations such as language, cultural influences were identified whilst going through the analysis. Overall through the information given in the following in this study more information regarding that warrants answers can be identified which will lead to proper care towards the teenagers who suffers from cystic fibrosis.

Introduction

Cystic fibrosis is an autosomal recessive disorder caused by specific gene variant inherited genetically from generation to generation. It impairs the clearance of mucus leading damages to the lungs and is the reason to other system dysfunctions like the digestive system and reproductive system. In the present world, total an estimated amount of 162,428 people are diagnosed with cystic fibrosis (CF) across 94 countries in the world. 65% are diagnosed and 12% are estimated to receive triple combinational therapy according to the registry data. It is found that approximately 1,000 new cystic fibrosis cases are found each and every year. CF is found in 2% of children, 40-50% of adults. Out of the numbers more than 75% of the population are diagnosed by the age of two. Today 19% of adolescents are estimated to be with CF which has shown a significant increase of teenagers with CF when compared between 1970 and 1982 as only 8.9% patients were adolescents with cystic fibrosis (Fitzpatrick, 1986).

Throughout the period of life, adolescence is considered as the most prominent as it is the time of rapid physical, cognitive and psychosocial growth and the extant of a chronic disease like cystic fibrosis could highly impact teenagers. Social disturbances or prevalence of anxiety and depression could be in teenagers with CF due to the health-guidelines and medications. However due to the severity of symptoms like wheezing, coughing or shortness of breath these factors are not been taken to proper considerations. It is stated that significant levels of emotional disturbances in adolescents have gone undetected in conventional studies (Carew, 2010).

Although the symptoms of the disease are being treated, another complication that can arise is anxiety and depression due to various reasons like decrease of life satisfaction, frequent hospitalization which leads to isolation from peer relationships (Besier, 2010). Suicide attempts is another factor that can be neglected besides anxiety and depression is needed for a solution to decrease or prevent them. One's own self-perspective is another maladjustment that the adolescents are struggling to adhere and overcome. Cystic fibrosis is a fatal disease that delays the pubertal growth in males and females. Teenagers who are keen to provide special care to one's own body or self-image can also be an emotional damage. Therefore, it is another factor that is warrants more attention because of cystic fibrosis on teenagers. Moreover since, occurrence rate of cystic fibrosis is recorded low in Asian countries, it is believed that no special care centers or facilities are being needed in those countries. This has led to a problem more CF cases have gone undetected and underreported. The rise of these inquiries clearly states that there is yet to find solutions or responses for the following, lack of findings and quantitative research is the main key for the scarcity of answers. It is also a field where more researchers are needed to be encouraged find answers.

This systematic review synthesizes the necessary gaps to explore the negative consequences of cystic fibrosis on teenagers by understanding the areas where more examinations and findings should be look into in order to provide better care for CF teenage patients.

Problem statement

Cystic fibrosis is a life-threatening disease with rather challenging symptoms to be faced, but due to these challenges all other impacts with regard to cystic fibrosis is being neglected. This shows that more findings and responses are needed to provide efficient to all adolescent CF patients. Although few studies have already been conducted about the aftermath of cystic fibrosis, there is only limited sources to prove and only paid attention to the population inside the European continents to grasp more attention of researchers to find possible solutions towards CF community. This review aims to identify the proper gaps that is needed to fill and warrant for answers with the hope of minimizing the impact of cystic fibrosis on teenagers.

Research questions

1. What are the major trends emerged from the systematic study of the effects of cystic fibrosis on adolescents?
2. What are the primary limitations and research gaps?
3. Which authors and journals have the most influence in this study?
4. Which keywords appear the most in this systematic review?
5. Who are the main contributors in this inclusive systematic analysis of how cystic fibrosis affect teenagers

Research objectives

1. To identify the main trends that are emerging from this systematic review the consequences of cystic fibrosis on teenagers
2. To examine the principle limitations and research gaps of this study
3. To discover the most influential authors and impactful journals based on this review
4. To analyze the most keyword occurrences in this systematic study
5. To identify the key author occurrences in this comprehensive analysis of effects of cystic fibrosis on teenagers

Methodology

The Preferred Reporting Items for Systematic Review was used as the methodology to approach the conclusion of this systematic review. In addition to that, in order to identify and understand the current research gaps this methodology contains evaluation and synthesis of research

findings. The purpose of this review is to analyze the existing research gaps and establish in impact of cystic fibrosis on adolescents.

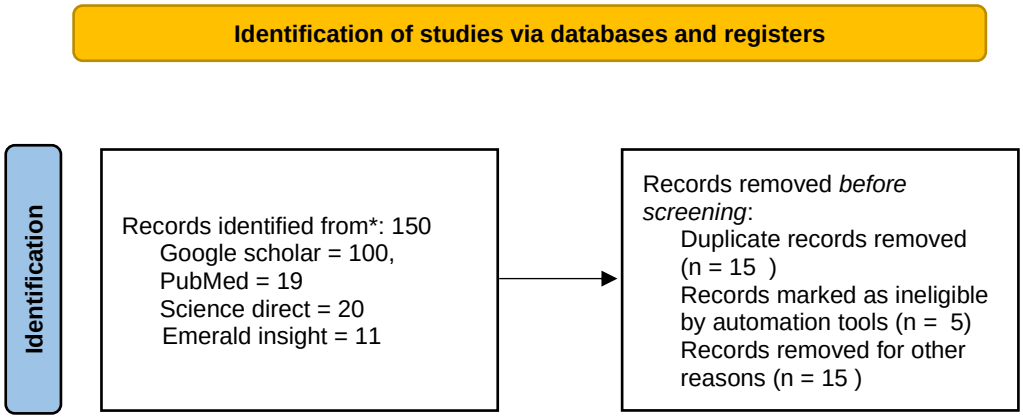
1. Qualifications of eligibility for the search

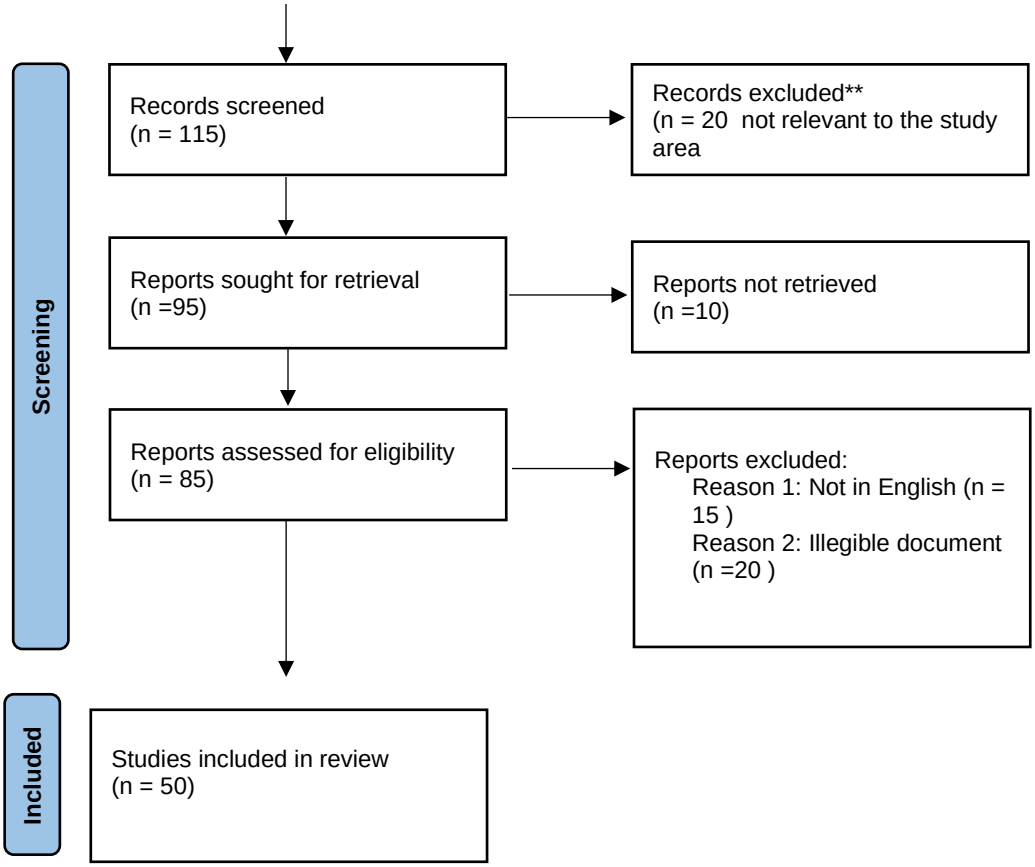
This review includes the most suitable studies that assess the social and mental impact of cystic fibrosis on adolescents. Furthermore, it has investigations regarding the cultural influence around cystic fibrosis. The findings gathered holds publications from 1976 to 2024. The assessments were conducted among adolescents. Databases that were used to find relevant research papers include Google scholar, PubMed, science direct and emerald insight. A total of 150 articles were selected initially for this systematic review. All searches were conducted without the inclusion of time frame in publication. A large synthesis of coverage which summarizes the existing knowledge and recognize gaps were from articles collected from the google scholar. The inclusion of key terms to find articles were “Cystic fibrosis”, “adolescents”, “Impact” “Teenager’s health” are the used Boolean in each database.

2. Clarifications of the search outcomes and completing the review analysis

This PRISMA 2020 flow diagram contains a total of 150 articles collected for this systematic review. 100 of the articles were found from google scholar, (19) was discovered from PubMed, (20) was taken from science direct and (11) from emerald insight. A total of 30 records were taken out for as they were duplicates and for other reasons. 5 records were also eliminated because they were records marked as ineligible by automation tools. 115 results were and 20 out of them was removed as they were not pertinent to the study area. 95 reports were sought for retrieval and 10 reports were not retrieved. After 85 reports were taken for evaluation 15 reports were excluded as they were not in English language and 20 were illegible documents. Therefore 50 studies are included for this review. The PRISMA 2020 standards for reporting systematic reviews and meta-analyses are adhered to in this systematic review.

Figure 1 - PRISMA chart





Growth (trend) of the publication

The growth or trend on the topic "Impact of cystic fibrosis on teenagers" has shown some amount of interest in the past couple of years. The diagram shows the noticeable shift of publications based on the impact of cystic fibrosis on certain years. From 1976 to 2002 small alterations of publications can be seen. However, after 2002 till 2008 there has been a peak for the analysis for the search of effects on cystic fibrosis. After 2008 there has been a significant of dip on the interest on performing researches about the review. But years between 2010 and 2014 appears to have another gradual increase of research papers. The significant fluctuating numbers is visible continuously after 2014 till 2018. More-over the expansion of undivided attention for the publications of the outcomes of cystic fibrosis can be seen in the diagram from 2021 until today. It can be concluded that there is an rise in the trend currently in the world to identify the consequences of cystic fibrosis on teenagers and suggesting the nurses and doctors in CF care-centres to be more aware of the situation.

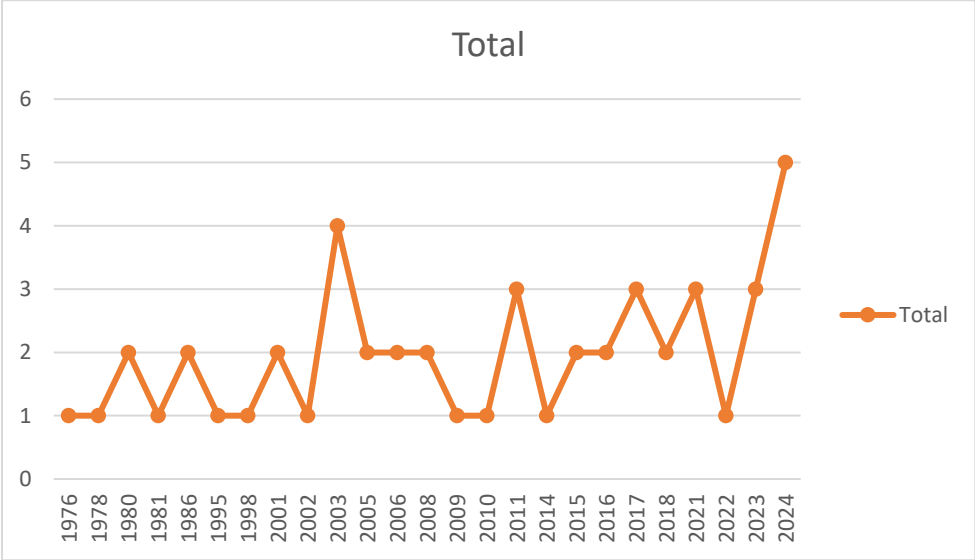


Figure 2- Growth or trend of the research from 1976 to 2024

Journals with the most influence of this study

H – index

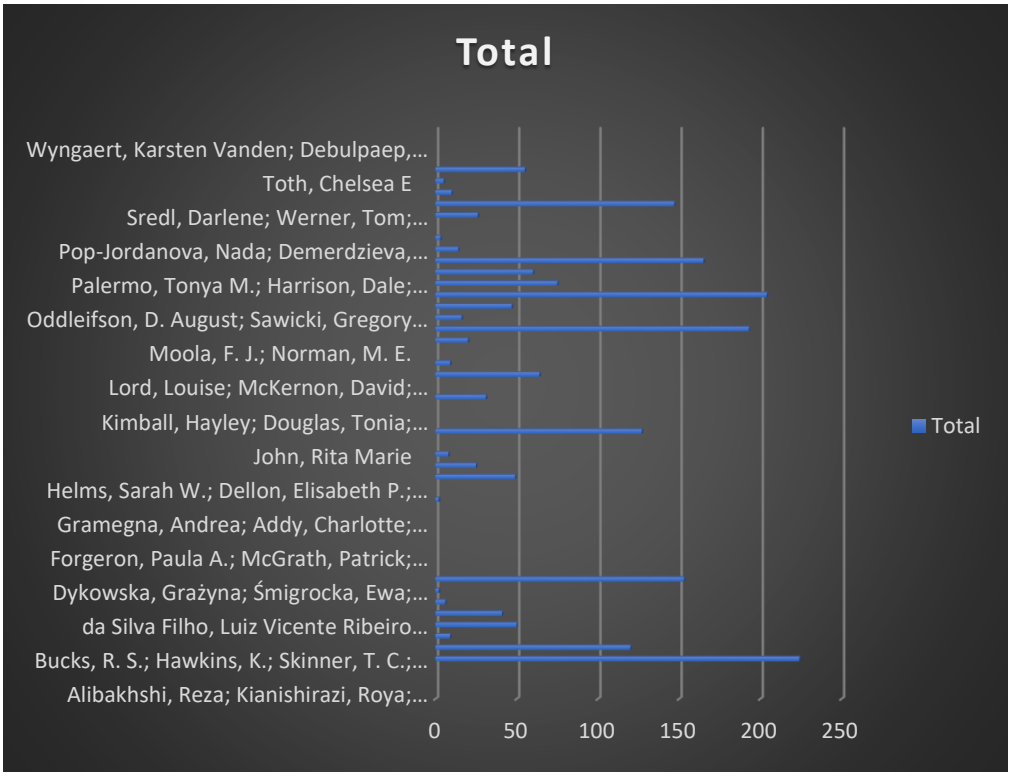
The information portal Recertify was used to categorize and find the main and the most applicable journals related to the study. Highest number of articles were gathered from The Journal of Cystic fibrosis which has an impact score of 4.63, a H-index of 91, a SJR 2.043 and an overall ranking of 1318. The number of articles obtained by this journal is 12. Another journal with total of three articles is the journal pediatric psychology which holds a impact score of 3.01, H – index of 139, a SJR of 1.143 and an overall ranking of 3564. Similar sum of articles has been contributed by Pediatric Pulmonology which accommodates an impact score of 2.43, it's H-index is 120, the SJR is 0.907 and an impact score of 5201. Another ideal journal that handed out a perfect article is the Journal of Pediatric Nursing which includes a impact score of 2.04, H – index of 74, a SJR 0.678 and scoring a overall ranking of 7773.

Journal	Impact score	H-index	SJR	Overall ranking
Journal of pediatric nursing	2.04	74	0.678	7773
Annals of American thoracic society	3.49	138	2.012	1350
Journal of interprofessional care	2.41	88	1.036	4187

Journal of cystic fibrosis	4.63	91	2.043	1318
Pediatric Respiratory reviews	3.97	75	1.42	2478
Journal pediatric psychology	3.01	139	1.143	3564
Child: Care, Health and development	2.33	98	0.725	7165
Journal of Clinical Medicine	3.07	113	0.882	5392
Journal of Youth and Adolescence	5.44	144	1.954	1404
Pediatric Pulmonology	2.43	120	0.907	5201

Most influential authors (citations)

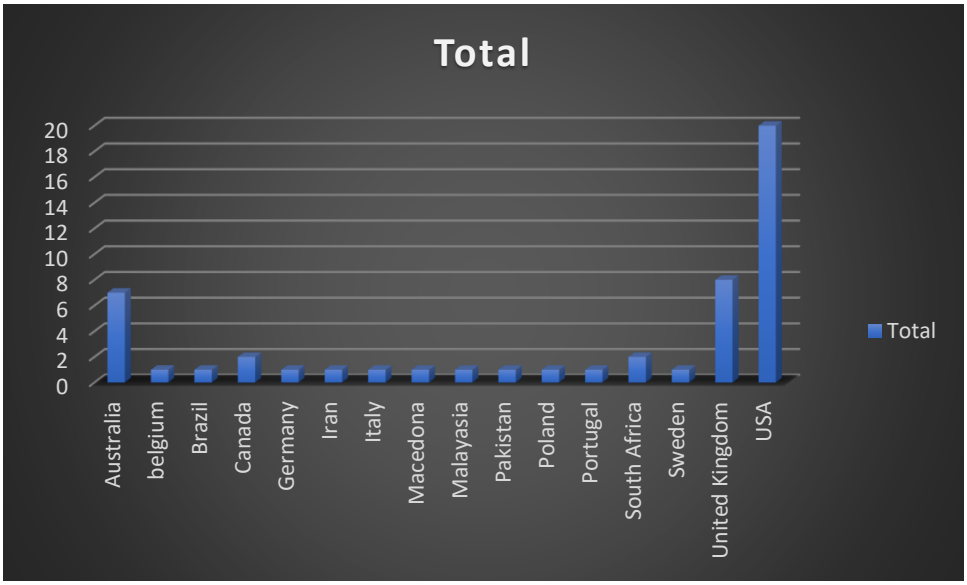
The below diagram depicts the most influential authors who has provided the publications based on the aftermath of cystic fibrosis on teenagers. One of the



prominent authors who has the publications Romola S bucks, alongside with Katharine Hawkins, T.C Skinner with a total sum of 224 citations. Another eminent author who has shown interest in the effects of cystic fibrosis is Richard W. Olmsted with few other authors like Ivy R. Boyle, Paul A. di' Sant Agnese and PH. D Sallyann Sack securing 204 citations. Ultimately, P.E Pfeffer also has a record number of citations 165 another formidable author has aroused attention for publications suggesting the warrant for a solution by the culminations of cystic fibrosis.

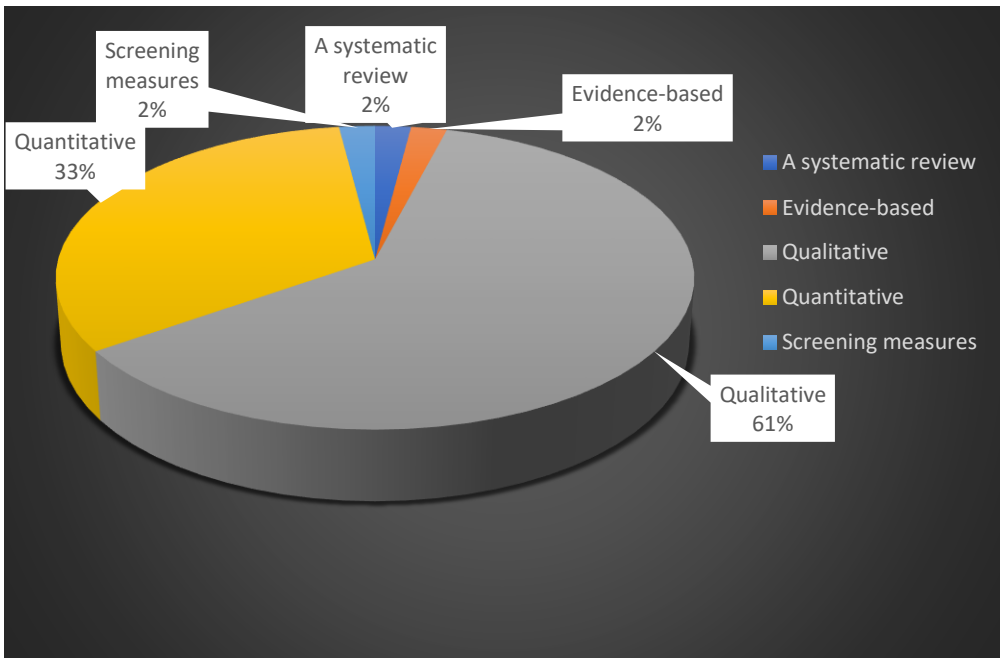
Countries of publications

The synthesis of countries with the most publications illustrates where the significant trend lies in the modern world. As stated by the information depicted in the graph United States holds the highest publications proving to be the frequent contributor to this research area with 20 publications. Furthermore, the next domain with most number of research findings is United-Kingdom contributing 8 notable findings is also known to provide their meaningful contribution to provide information. Australia falls third proving to show the similar inquiry like the previous countries by supplying 7 publications under the relevant research area. In addition to that, other countries that shows their involvement by participating and providing their factors are Canada, South-Africa, Belgium and Germany. Though there are less acceptable articles that falls under the topic, these few domains were have provided excellent details which align with this field of study.



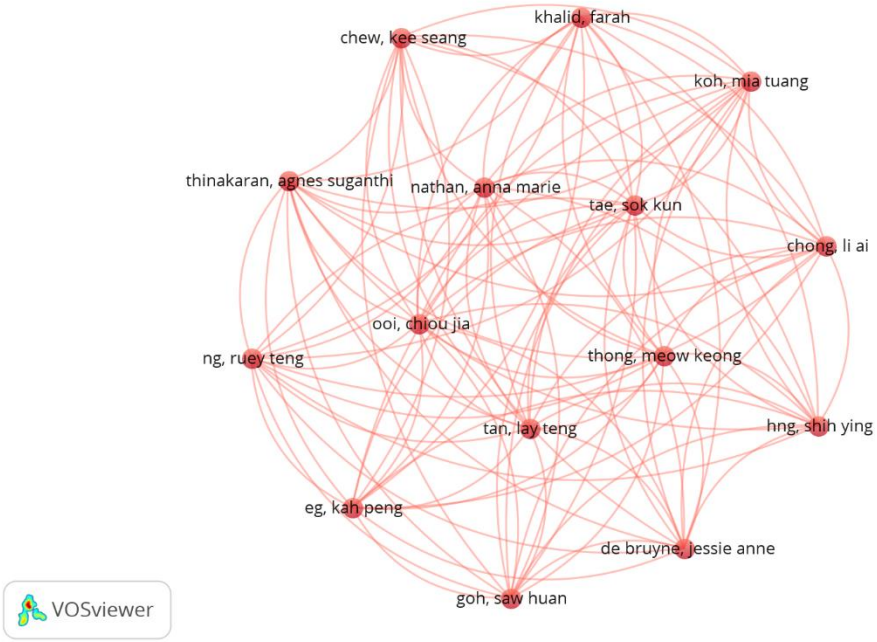
Research techniques used in this review

Results of studies on consequences of cystic fibrosis have been approached in various types of methods. Qualitative method is the most used methodology to gain information based on the field of research. 61% of qualitative methodology is being utilized. More over the next often used methodology falls under the classification quantitative method. 33% (16) studies are found through the method quantitative studies making the second most involved method to find data regarding this field of study. Evidence-based and screening measures are two alternative techniques applied. These strategies gives a clear visualization of the factors that is requires more attention to improve the patient care for the CF adolescents in the teenage community.



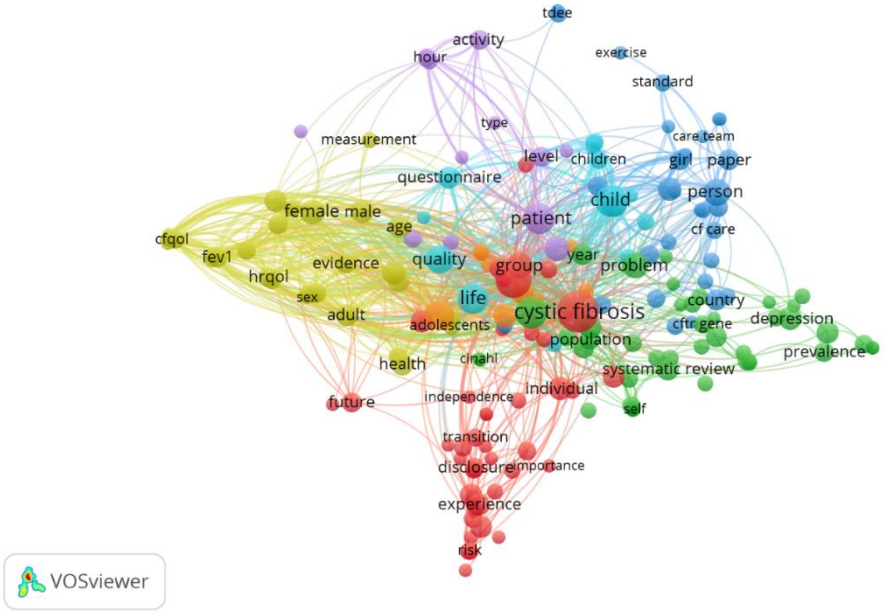
Analysis of co-author occurrences

The visualization of co-authorship network offers important insights into the dynamics of a collaboration within this field of study. Especially, authors such as, Thinakaran Agnes Suganthi, Nathan Anne Marie, de Bruyne Jessie Anne, Thong Meow Kong and other respective writers confirms to be the most impactful inter-connected authors in this research domain. Even though, the diagram states that the arrangement of authors densely connected, it can also be possible verify that the engagement of more authors seems low. Therefore, it can be confirmed that this visualization is an effective tool to understand the need of involving more authors to find the negative repercussions of cystic fibrosis in adolescents.



Analysis of keyword co-occurrences

The visualization of keyword co-occurrences network provides thorough look into the research environment regarding the impact of cystic fibrosis on adolescents. In the center of diagram points out the primary keyword “cystic fibrosis” that highlights main significance of this research area. Furthermore, the keywords around initial keyword like “adolescents”, “quality”, “life” represents the broad concepts that run throughout the work. Each cluster in the given represents important areas related to the study. The clusters emphasize who are being affected and what areas are affected in adolescents who suffers from cystic fibrosis by illustrating the breadth of existing knowledge and offering possible lines of inquiry. Therefore, this illustration can be considered as a powerful tool for all researchers to identify the necessary that requires immediate attention.



Discussion

This systematic review supplies a thorough analysis to illuminate the complicated impact of the cystic fibrosis on adolescents. This study has summarized the existing knowledge, identify the gaps and seeks to provide proper revelations to enhance the patient care given to adolescents in the CF community. Moreover, its objective is to provide capitative information to all future researchers and mainly the health-care teams working in CF care centers.

The standard of life and survival rate of CF patients have increased due to the technological advancements, thus evolving CF from being child-hood terminal disease to a chronic disease making new born babies with cystic fibrosis transition into adolescents. However, through the alterations of children into adolescents have created the context of teenagers facing health-related stigma. Moreover, interferences with social interactions among teenagers have also proved to increase the influence of the stigma. Therefore, it is important to verify the psychological that influence CF adolescents **(Oliver, 2014**).

Another disastrous difficulty faced by teenagers with CF is prevalence of depression and anxiety. Whilst adolescence is considered a pivotal stage in life, the effects of cystic fibrosis lead to teenagers with anxiety. Through frequent hospitalization isolation from peers can arise. Not only that but also by the consumption of high caloric food in order to maintain the nutrition, their desire to maintain a proper body figure is being utterly disrupted. Hence leading to anxiety and depression among the CF community. However, information regarding mental health problems with relate to CF is confined is being overlooked (**Besier, 2011**). In addition to that, another result that affects the psychological outcomes is medical mistrust. This has shown signs of poorer pulmonary function test results and adherence, and other health consequences (**Kow, 2023)**

Cystic fibrosis causes thick mucus in exocrine glands resulting in obstruction of the pancreas. This causes to delayed pubertal growth and sexual development. CF males showed signs of unusual adjustments to compared to disturbed males with disturbed males and pubertal and growth delayed males. Hence proving that male teenagers with CF has adjustments issues about their self-perception because of the signs and symptoms caused by cystic fibrosis. This factor is warrants more lime-light nutrition in teenagers **(Landon, 1980).** There is a limitation of intervention programs that test efficacy to enhance self-perception or self-concept (Harrigan,2024).

Over the years, cystic fibrosis has been known as a rare occurrence outside Caucasian population. From the cultural influence regarding cystic fibrosis it is been stated that screening process for cystic fibrosis should not be considered in populations outside the Europe. Nevertheless, this overlooked factor has created doubts whether cystic fibrosis reports are being misdiagnosed with other diseases like tuberculosis, malnutrition or being under-reported. Even though a smaller number of cases are identified in Asians, a review of screening methods should be conducted to capture more incident rates and provide more attention towards the possible challenges (Duff, 2003).

Limitations identified from the Review

It is essential to acknowledge the limitations found in the review. One of the limitations was the methodological weakness in order to identify the necessary issues regarding the problems related to the impact of cystic fibrosis. More intervention programs are needed to identify the psychological risk factors which affect the adolescents.

Moreover, it is clear that some studies are being overlooked due to complexity of the disease outcomes, therefore important facts are being missed to provide patient care to the adolescences in the CF community. Furthermore, socio-cultural challenges have also become a difficulty creating geographical boundary that more research of this domain is done in countries like USA, United Kingdom and countries in the European region. Furthermore, the prejudice in the literature review is also caused due to the language issue as publications in English language

are taken into considerations while publications which has more and clear factors regarding the issue is being published in other languages.

Research gaps identified in the study

Methodological gap - Even though the utilization of quantitative methods are used to identify the gap, there is a still quantitative gap as only 33% of quantitative methods are being used. There is a space to fill in research findings with more mixed methodologies by combining more qualitative and quantitative methods. More SLRs are also needed for this field of study. This will be beneficial to identify more gaps accurately and effectively.

Empirical gap – The examination of the contributed publications shows that more evaluation and verification is highly needed in the impact of cystic fibrosis on teenagers mentally and socially besides the complexity of the disease basis. Due to the cultural influence or stigma less publications exist in the Asian region when compared to United states of America, United kingdom, Canada or Australia. By combining these gaps will liquidity quality patient care to all adolescents in the CF community and reduce the impacts of cystic fibrosis.

Future research directions

Conducting more research based on the psychological factors and the social and cultural issues related to cystic fibrosis is a must to look into as a solution. As for more future guidance for the researchers, more reports of diagnosis of cystic fibrosis should be included in the Asian and region implementation of diagnosis test, screening methods.

An ideal understanding about the negative effects caused to the psychological side of all CF teenage patients should be given to all cystic fibrosis health-care team.

By enhancing more methodologies like systematic reviews would help to identify more gaps and unanswered questions. In addition to that, more mixed methodologies should also be conducted. By bridging more quantitative and qualitative data understanding the comprehensive context regarding the cystic fibrosis can be a beneficial outcome for all teenagers.

Conclusion

The above data depicts that it is essential to understand and identify the areas that is needed to be filled with answers regarding the effects of cystic fibrosis on adolescents. More research is needed to refine the prevalence of mental health disturbances like anxiety and depression which could lead to diagnostic confusions (Drotar, 1978).

These findings encapsulate cultural issues regarding the disease faced by the patients. The belief of the cystic fibrosis being non-existent outside European populations due to low occurrence rate of CF and not holding screening programs. This results in under-reports and misdiagnosis with other complications like chronic pulmonary infections. Yet reports have stated that there are different variants of CFTR found in Iranian patients (Alibaskshi, 2008). Hence proving that cystic fibrosis now is major issue around the world.

Moreover, the social stigma, maladjustment of all adolescents by the disease complications and issues regarding the self-perception was also reviewed in this study. It was concluded there is a methodological gap as more mixed methodologies should be conducted to again more clear comprehensive analysis regarding the issues faced by cystic fibrosis. In addition to that, more quantitative analysis should also be conducted alongside with systematic reviews.

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