REVIEW ARTICLE

CYSTIC FIBROSIS-ASSOCIATED LIVER DISEASE: A REVIEW ARTICLE

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ABSTRACT

Cystic fibrosis-associated liver disease (CFLD) is animportant aspect of cystic fibrosis (CF), which manifests with various signs and symptoms. Lack of specificexaminations for CFLD have made the diagnostic process of the disease time-consuming, and the disease is often identified after severe progress. Finding the associations between the outcomes of various clinical,biochemical, and sonography examinations could help specialists identify the disease in a timely manner. This review study aimed to determine the correlations between the outcomes of various diagnostic approaches based on the current literature. According to the literature, some studies have reported correlations between various diagnostic approaches, while other studies have reported no associations in this regard. This discrepancy could be due to the various manifestations associated with CF..

INTRODUCTION

Cystic fibrosis (CF) is the nearly prevalent life-shortening, autosomal, recessive disease in a number of areas across the world, such as the United States, Europe, and Australia. The incidence of CF has been reported to be 1:3,500 live births [1,2]. Currently, the life expectancy of CF patients has bigger to further than 40 living in developing countries owed to the increase in the management of the disease [3]. However, CF is nevertheless coupled with compound complications, such as hepatic cirrhosis, pulmonary failure, diabetes, and osteoporosis. amongst different complications of CF, liver disease is measured of great magnitude anticipated to its climax commonness and lay bare of mortality [4]. According to the definition by North America, cystic fibrosis-associated liver disease (CFLD) is indicated by the presence of liver cirrhosis and portal hypertension, persistent elevated liver enzymes, fibrosis, steatosis, and abnormal changes in ultrasound patterns [5,6]. The comprehensive global prevalence of CFLD has been reported to be 37.9%. According to statistics, approximately 2.5% of the global mortality in the patients with CF is connected with liver disease, which is notorious as the third leading cause of loss of life in these patients [7].

To date, liver biopsy has been the nearly gold

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standard technique for the assessment of CFLD. However, this technique enveloping and irregularly leads to difficult complications in the patients [8]. Among a choice of diagnostic methods, non-invasive techniques for the diagnosis and management of CFLD control attracted the concentration of specialists. Furthermore, early diagnosis of CFLD is of paramount importance due to the possible positive effects of ursodeoxycholic acid therapy, and there is an urgent need for the development of the trial of its prophylactic use. Some of the routine procedures for the diagnosis of CFLD include determining the clinical characteristics, biochemical tests, and sonography. Evidently, finding a rational correlation between various assessments could further support the early detection of CFLD.
This review study aimed to present the results of previous studies regarding the correlations between the outcomes of various diagnostic approaches for CFLD.

LITERATURE REVIEW

Clinical Features of CFLD

Detection of CFLD is challenging since it is a lot a subclinical disease and manifests with a broad form of signs and symptoms. The clinical appearance of CFLD might refrain from specialists to suspicious the disease and prescribe additional examinations for the patients in succession to bear out the diagnosis (10). In a study, Lamireau et al. exhibit demonstrated that CFLD is further shared in younger family (prevalence: 41%) at the period of 12 years. Similarly, Colombo et al. reported that the incidence time of liver disease is top in the patients with the history of meconium ileus, male patients, and individuals with severe genotypes .

In genotypes, factors such as pancreatic insufficiency and severity of pulmonary disease come up with been reported to begin CFLD. In a analysis conducted on 288 patients with CF, 256 had pancreatic insufficiency, and approximately one-third of these patients (n=80) were diagnosed with liver disease. some studies well described the other manifestations of CFLD, including portal hypertension, neonatal cholestasis steatosis, elevation of liver transaminases, lack of alkalization, and bile dehydration.

In this regard, the findings of Corbett indicated growth retardation and poor nutrition status importance in the patients with CFLD (13). Furthermore, factors such as the history of jaundice, changes in thestool pattern/ colour, abdominal pain, loss of weight, and family history of liver diseases must be careful in the patients with high levels of liver aminotransferases who are the possibility family history of liver diseases [12]. According to the examine by Ciuca et al., evaluation for portal hypertension, liver cirrhosis, and pancreatic insufficiency supposed to be measured as the first assessment of CFLD.

Diagnosis of CFLD

Biochemical Test

The serum assessment of transaminases is the most common test performed for the diagnosis of liver disease. However, the distance from the ground of liver enzyme levels is everyday in the patients with CF, therefore that it would not be linked with strict liver disease in every single one the cases.

Liver function test (LFT) was investigated in a group prospective study in this regard. According to the obtained results, approximately

25% of the patients had abnormal LFT, and in single 13% of the cases, a large overtone was experiential between high-minded LFT and clinical outcomes .

In a further research, Lindblad et al. assessed the pathological morphology of liver biopsies in quite a lot of patients, and barely a slender rise was practical in their serum transaminase levels . On the other hand, a variety of consider cover indicated that children with CF, who were exaggerated by multilobular biliary cirrhosis and rigorous fibrosis, had typical elevation of liver enzymes .

In a another research conducted by Williams during a nine-year period, the authors reported that the consequences of biomarker tests differed from sonography findings, and no correlations were experiential in this regard. In addition, in 3% of the patients with persisting abnormalities of the liver echo surface and persisting splenomegaly, the intensity of aspartate aminotransferase were inside the common range. In the mentioned study, 725 ultrasound examinations were performed .

Ling et al. followed-up 124 patients with CF for the substantiation of liver disease for four existence [17]. According to the findings, 40% of the patients with abnormal clinical or ultrasound examinations had biochemically augmented aminotransferase levels. During the stick to up, 48% of the patients were practical to cover liver abnormalities in the clinical, biochemical, and ultrasound examinations. several of the before studies in this observe state furthermore definite the correlation between ultrasonographic findings in the patients with CF with their clinical and biochemical characteristics [18]. Overall, Ling concluded that ultrasound and the clinical examination of abnormalities based on biochemical tests might outcome in the induce identification of CF in the patients .

ULTRASONOGRAPHY

Ultrasound is a non-invasive, cost-effective, and exceedingly useful procedure for the diagnosis of hepatic steatosis, cirrhosis, and the complications caused by portal hypertension (e.g., ascites and splenomegaly). However, ultrasound cannot reliably rule out early on liver disease [20].

According to the study by Leung et al., sonography abnormalities were practical in 18% of the patients with pancreatic scarcity in CF. The findings of the mentioned examination are coherent with the contemporary copy in this regard. even though 3.3% of the patients with no data of liver disease (e.g., portal hypertension and thrombocytopenia) had cirrhosis based on their ultrasound results. This may well be exactly to the manifestation of cirrhosis initial in life. Furthermore, no correlations were reported between meconium ileus, malnutrition, deteriorated FEV1, and ultrasound results. However, Leung claimed that meconium ileus is the likely lay bare issue for a homoge.

an alternative investigate in this have to do with was conducted on an infant population, and the consequences demonstrated no major disparity between meconium ileus and enhancement of liver disease in the patients with CF [22]. Moreover, Colombo investigated 177 patients with CF with a 14-year follow-up. According to the consequences of the mentioned study, the ultrasonographic patterns of 10% of the gear showed the presence of cirrhosis, bit near was

no sign of portal hypertensions .

In another study performed on 174 patients with CF, completely were followed-up every day for clinical, biochemical, and ultrasonography results. According to the findings, three children's develops CFLD during infancy, all along with the signs of portal hypertension. The outcome of ultrasonography also confirmed the progression of portal hypertension. According to the literature, multilobular cirrhosis with the developmentof portal hypertension is a most important liver disease in the patients with CF .

CONCLUSION

liver abnormalities has been reported in several of the patients with CF with normal biomarkers, as many studies be inflicted with demonstrated that the improper functions of the liver was evident in the clinical, biochemical, and ultrasound assessments of these patients. The discrepancies between the findings may possibly be owed to the varied assortment of liver disease manifestations, which may have an advantage to a variety of fallout in unusual investigations.

With respect to ultrasonography, a correlation has been reported between ultrasound patterns and clinical data in some studies, while other studies have denoted no such association. In fact, the correlations of various clinical symptoms have been reported variably in different studies. To obtain better results, it is suggested that specific research be conducted independently on the prevalent types of liver diseases to determine the correlations between various assessments.

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CONFLICT OF INCIDENCE

No conflict of interest declared by the authers.

AUTHORS' CONTRIBUTION

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