بسم الله الرحمن الرحيم

MONA MAGHRABY

شبكة المعلومات الجامعية

التوثيق الالكتروني والميكروفيلم

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جامعة عين شمس

التوثيق الإلكتروني والميكروفيلم

قسم

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علي هذه الأقراص المدمجة قد أعدت دون أية تغيرات

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تحفظ هذه الأقراص المدمجة بعيدا عن الغبار

MONA MAGHRABY

Liver involvement in children with cystic fibrosis

Thesis

For Partial Fulfillment of Master Degree
in Pediatrics

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بسم الله الرحمن الرحيم

قالوا

سبحانك لا علم لنا
إلا ما علمتنا إنك أنت
العليم الحكيم

صدق الله العظيم

سورة البقرة الآية: ٣٢

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Mohamed Magdy Saleh Al Emam

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List of Abbreviations

Abb.	Full term
ABC	*ATP-binding cassette*
ALP	*Alkaline phosphatase*
ALT	*Alanine aminotransferase*
AST	*Aspartate aminotransferase*
CBC	*Complete blood count*
CF	*Cystic fibrosis*
CFF	*CF Foundation*
CFLD	*Cystic fibrosis associated liver disease*
CFRD	*Cystic fibrosis related diabetes*
CFTR	*Cystic fibrosis transmembrane conductance regulator*
CRMS	*CFTR-related metabolic syndrome*
DIOS	*Distal intestinal obstruction syndrome*
GERD	*Gastroesophageal reflux disease*
GGT	*Gammaglutamyltransferase*
IBD	*Inflammatory bowel disease*
IRT	*Immunoreactive trypsinogen*
NAFLD	*Non-Alcoholic Fatty Liver Disease*
PERT	*Pancreatic enzyme replacement therapy*
PSC	*Primary Sclerosing Cholangitis*
SIBO	*Small intestine bacterial overgrowth*
SSC	*Secondary Sclerosing Cholangitis*
UDCA	*Ursodeoxycholic acid*
US	*Ultrasonography*

INTRODUCTION

Cystic fibrosis (CF) is a systemic disease, of autosomal recessive inheritance in the course of which pathologies of different systems and organs can be observed. The severity of the disease, and the survival time of patients, is determined by the degree of changes in the respiratory system **(Kobelska-Dubiel et al., 2014).**

Due to progress in the treatment of cystic fibrosis, the life span of such patients has been prolonged. This can lead to a display of pathologies of other systems and organs that were not previously observed in this disease due to the short duration of the patient`s life **(Kobelska-Dubiel et al., 2014).**

Cystic fibrosis associated liver disease (CFLD) belongs to the group of common symptoms of this disease; however,due to the lack of specific and sensitive CFLD diagnostic markers, the epidemiological data may be incomplete **(Siano et al., 2010).**

According to various sourses, the prevalence rate of CFLD,diagnosed on the basis of clinical, biochemical and imaging (ultrasonography) tests, is 2–37% in children and young adults **(Wilschanski et al., 2011).**

Currently it is believed that CFLD is the third most frequent cause of death in patients with cystic fibrosis, after lung disease and complications related to organ transplantation, it constitutes 2-4% of CF cases **(Minagawa et al., 2007).**

The clinical picture of CFLD can take many forms, from mild asymptomatic hypertransaminasaemia to cirrhosis in infants with cystic fibrosis, liver dysfunction may occur in the form of cholestasis. Cholestasis in infants occurs relatively rare; in about 50% of cases, it co-occurs with meconium ileus and parenteral nutrition **(Minagawa et al., 2007).**

The pathomechanism of this phenomenon is the thickening processes of acidophilic secretion in the intrahepatic bile ducts. The clinically significant pathology of the hepatobiliary system in patients with cystic fibrosis is characterised by a slow but progressive clinical course **(Herrmann et al., 2010).**

The CFLD diagnosis may cause many difficulties as a "gold standard test" hasn`t yet to be developed. Early diagnosis of CFLD is very important because clinical symptoms appear late, when hepatobiliary system damage is already very advanced. Some studies suggest that only during the early stages are histopathological changes reversible and may be efficaciously treated **(Siano et al., 2010).**

AIM OF THE WORK

The main objective of the study is:

To search for different forms of liver involvement in children with cystic fibrosis and to detect predictive factors for liver affection in those children.