بسم الله الرحمن الرحيم

MONA MAGHRABY

شبكة المعلومات الجامعية

التوثيق الالكتروني والميكروفيلم

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جامعة عين شمس

التوثيق الإلكتروني والميكروفيلم

قسم

نقسم بالله العظيم أن المادة التي تم توثيقها وتسجيلها
علي هذه الأقراص المدمجة قد أعدت دون أية تغيرات

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تحفظ هذه الأقراص المدمجة بعيدا عن الغبار

MONA MAGHRABY

Sputum Neutrophil elastase in pediatric bronchiectasis

Thesis

Submitted for partial fulfillment of master's degree in pediatrics

By

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Candidate

✍ ***Heba Hossam El-Din Mohamed Abd-Elhak Radwan***

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Abb.		Full Term
ABPA	:	Allergic bronchopulmonary aspergillosis
ACOG	:	American College of Obstetricians and Gynecologists
AHR	:	Airway hyper-responsiveness
ANOVA	:	Analysis of variance
ATS	:	American thoracic society
BAFF	:	B-cell activating factor
BAL	:	bronchoalveolar lavage
BALF	:	Broncho Alveolar Lavage Fluid
BMI	:	body mass index
BSI	:	bronchectasis severity index
C1	:	Complement 1
c3b	:	component 3 b
C5a	:	complement 5
CAMP	:	Cyclic adenosine monophosphate
C-ANCA	:	antineutrophil cytoplasmic antibodies
CCL2	:	Chemokine Ligand 2
CCL23	:	Chemokine ligand 23
CCL3	:	Chemokine Ligand 3
CCL4	:	Chemokine Ligand 4
CF	:	Cystic fibrosis
CFB	:	cystic fibrosis bronchiectasis
CFLD	:	CF-associated Liver Disease.
CFTR	:	Cystic fibrosis transmembrane conductance regulatoR
COPD	:	Chronic obstructive pulmonary disease
CT	:	Computerized Tomography
CXCL1	:	The chemokine ligand 1
CXCL10	:	chemokine ligand 10
CXCL8	:	Chemokine Ligand 8
CXR	:	Chest x-rays
DIOS	:	distal Intestinal Obstruction Syndrome
ENAC	:	Epithelial sodium channels
ERS	:	European respiratory society
ESCF	:	Epidemiologic Study of Cystic Fibrosis
F508DEL-CFTR	:	cystic fibrosis transmembrane conductance regulator

FASL	**:**	Fas ligand
FEV1	**:**	forced expiratory volume.
FVC	**:**	forced vital capacity
GC	**:**	general condition gonococcus
G-CSF	**:**	Granulocyte colony-stimulating factor
GI	**:**	Gastrointestinal
HB-EGF	**:**	Heparin-binding EGF-like growth factor
HCO3	**:**	Bicarbonate
HRCT	**:**	High Resolution Computed Tomography
HS	**:**	hypertonic saline
ICS	**:**	Inhaled corticosteroids
ICU	**:**	intensive care unit
IL-1ra	**:**	interleukin-1 receptor antagonist
IL-6	**:**	Interleukin 6
IL-8	**:**	interleukin 8
IL-B	**:**	interleukin beta
IV	**:**	Intravenous
KB	**:**	Keratodermia blennorrhagicum
LDLT	**:**	living donor liver transplantation
MI	**:**	myocardial infarction
MMP-8	**:**	matrix metalloproteinase 8
MMP-9	**:**	matrix metalloproteinase 9
MPO	**:**	Myeloperoxidase
MUC5AC	**:**	Mucin 5AC
MUC5B	**:**	Mucin 5B
NACL	**:**	sodium chloride
NCFB	**:**	Non-cystic fibrosis bronchiectasis
NE	**:**	neutrophil elastase
NETS	**:**	Neutrophil extracellular traps
NIP	**:**	National Immunization Program
PBS	**:**	Phosphate-buffered saline
PCD	**:**	Primary ciliary dyskinesia
PERT	**:**	Pancreatic Enzyme Replacement Therapy
PFT	**:**	pulmonary function tests
PH	**:**	potential hydrogen
PMN	**:**	Polymorphonuclear leukocytes
PTC	**:**	Percutaneous transhepatic cholangiography

QOL	**:**	quality of life
SMC	**:**	structural maintenance of chromosomes
TNF ALPHA	**:**	Tumor necrosis factor alpha
TRAIL	**:**	TNF-related apoptosis-inducing ligand
VEGF	**:**	Vascular endothelial growth factor
VX-770	**:**	Lipophilicity of the Cystic Fibrosis Drug, Ivacaftor
WHO	**:**	World Health Organization.
α1-AT	**:**	Alpha-1 antitrypsin
α1-AT	**:**	alpha 2 anti-trypsin

INTRODUCTION

Bronchiectasis is a chronic, debilitating respiratory condition that affects people of all ages. The condition is characterized by a vicious cycle of persistent bacterial infection and excessive neutrophilic inflammation owing to impairment of airway defense mechanisms. Patients have daily excessive sputum and associated symptoms, recurrent chest infections and impaired health-related quality of life **(Smith et al., 2017)**

Cystic fibrosis is caused by a gene mutation leading to dysfunction of the cystic fibrosis transmembrane conductance regulator (CFTR) protein. It affects multiple organ systems—the lungs, pancreas, upper airways, liver, intestine, and reproductive organs—to varying degrees. Its diagnosis requires both clinical evidence (positive newborn screening, sibling[s] with cystic fibrosis, clinical signs) and the demonstration of CFTR dysfunction by an elevated chloride concentration in sweat, and/or two disease-causing mutations, and/or abnormal electrophysiological findings (nasal potential difference measurement, intestinal short-circuit current measurement) (**Naehrig et al.,2017)**

Human neutrophil elastase (NE) is a proteolytic enzyme belonging to the chymotrypsin-like family of the serine-proteinases. NE is a 218 amino acid long protein packaged in