Cystic fibrosis medical terminology

Updated April 2018

A

Airway Surface Liquid – a thin film of liquid that lines the airways

Anabolic Steroids – a synthetic male sex hormone that promotes weight gain

Aspergillosis – infection by the fungus Aspergillus fumigatus. It is one of the most common Aspergillus species to cause disease in individuals with an immunodeficiency. It is typically found in soil and decaying organic matter, such as compost heaps

Azithromycin – an antibiotic which benefits patients with chronic Pseudomonas aeruginosa

B

Biofilm – a community of microorganisms attached to a sold surface

Bronchoscopy – a procedure to allow your doctor to look at your airways with a thin viewing instrument

Burkholderia cepacia – bacteria found in soil and other items such as decaying onions. Infection can cause serious illness in people with CF

Cachexia – abnormal weight loss and weakness associated with a serious illness

Cirrhosis – fibrous change in the liver in response to things like infection or obstruction of bile flow

Compressor – electric pump used to power nebuliser for inhaled delivery of medicine

Congenital – something you are born with

Cross infection – the transmission of bacteria and infections from one person to another

Creons – enzymes taken with food to help absorb nutrients and vitamins

Cystic fibrosis conductance regulator (CFTR) – a gene that codes for a protein that conducts chloride across the membrane of airway cells. Defaults of the CFTR gene affecting chloride ion channel function lead to problems with fluid transport in the lung, pancreas and other organs, resulting in cystic fibrosis

Cystic Fibrosis Related Diabetes (CFRD) – Diabetes specifically caused by cystic fibrosis. CFRD develops with age

D

Domino transplant – the heart of the CF recipient of a heart-lung transplant is donated to another patient who requires heart transplant

Duodenum – the first part of the upper small bowel where the pancreatic and bile ducts enter

E

Enzyme – a protein that speeds up biological reactions

Exacerbation – an increase in severity of a condition – often used to describe a drop in the health of the lungs

F

F508del – this is the most common cystic fibrosis gene. There are over 2000 different genes related to cystic fibrosis.

G

Gene – a piece of DNA that contains all the necessary information to manufacture a protein

Genome- the total genetic material of an organism and person

Genotype – the genetic makeup of an individual

H

Heterozygote – a healthy person carrying one faulty gene such as CF. This term is also used in research to describe some who has two different CF genes i.e. they could heterozygote for the common F508del gene which means they also carry another different CF gene.

Homozygote – a person with two copies of a faulty gene and who has cystic fibrosis.

I

Ibuprofen – non-steroidal anti-inflammatory used to suppress inflammation in the lungs

Ileum – the lower part of the small intestine.

Immune suppressed – decreased resistance to infection due to having lower immunity to infections

Intubation – insertion of a tube into the airways

J

Jejunal biopsy – obtaining tiny pieces of the lining of the upper bowel to examine with a microscope

L

Laparotomy – surgical cut into the abdominal cavity

Lung transplant – a surgical operation in which a patient's unhealthy lungs are replaced by lungs which come from a donor

M

Malabsorption – failure to absorb nutrients, usually referred to with the bowel

Meconium ileus – intestinal obstruction in newborns with CF

Meliodosis – a disease carried by wild rodents transmissible to people and can cause fatal blood poisoning, pneumonia and abscesses. It is a risk in some tropical climates

Metaneb – a hospital-based therapy which combines three treatments in one – volume expansion, secretion clearance and nebuliser therapy.

Mucolytic – a substance or technique that thins mucus

Mucus – slimy substances secreted by mucous membranes

Mycobacterium – bacteria of the same group that causes tuberculosis.

N

Nasal potential difference – electrical potential across the lining of the nose or airways and affects people with CF

Nebuliser – device/equipment used for inhaling a liquid medicine such as hypertonic saline or antibiotics

Nonsense mutation – a point mutation in a sequence of DNA that results in a premature stop codon. Responsible for about 10 percent of cystic fibrosis cases worldwide

Nontuberculosis mycobacterium (NTM) – an infection that causes lung disease that resembles tuberculosis

O

Oedema – excessive accumulation of fluid in the body

P

Paediatrician – a doctor who specialises in medical treatment of children

Pancreas – a gland that lies behind the stomach and secretes digestive enzymes into the upper bowel and insulin into the blood

Pancreatic sufficient – a person with enough pancreatic function to absorb fat normally

Pathogen – a microscopic organism that causes disease

PIC line – a peripherally inserted central catheter (PIC line) is a form of intravenous access that can be used for a prolonged period of time e.g. extended antibiotic therapy.

Polyp – a growth protruding from a mucous membrane – such as the lining of nose or bowel

Port-a-Cath (Port) - is a small medical appliance that is inserted beneath the skin. It is attached to a catheter used to give intravenous fluids

Pre-implantation Genetic Diagnosis (PGD) – also known Pre-Natal Diagnosis (PND), this enables two cystic fibrosis carriers to have a child that does not have cystic fibrosis. IVF is used to conceive a baby and then the embryos screened for cystic fibrosis before implantation

Pseudomonas aeruginosa – a major cause of chronic chest infection in CF

Pulmonary Specialist – a medical doctor who specialises in lung and breathing disorders

R

Respiratory Specialist – a medical doctor who specialises in lung and breathing disorders

S

Spirometer – an instrument for measuring the amount of air inhaled and exhaled

Sputum test – a test used to detect and identify bacteria or fungi that infects the lungs or airways

Staphylococcus aureus – a common germ that infects the airways of people with CF

Stoma – an opening into a hollow organ

T

Tobramycin – antibiotic prescribed to treat Pseudomonas

Trachea – main wind pipe

Trypsin – pancreatic enzyme that digests protein

U

Unique strain – a strain of bacteria or bug that is only found in that patient – often applied to Pseudomonas