



# Respiratory - Pulmonary System

The main function of the respiratory system (also known as the pulmonary system) is to supply the body with oxygen and rid it of carbon dioxide. This process also removes metabolic wastes and maintains the pH balances of the body. The organs involved are the airways, lungs, and muscles that mediate the movement of air into and out of the body. When you breathe, air comes in through the mouth and nose. It passes through the larynx, also known as the voice box, and enters the chest cavity through the trachea. The trachea then splits into two tubes called bronchi, each of which leads into a lung. The airways become progressively smaller and at the end are small sacs known as alveoli, which are surrounded by capillaries. The oxygen from the air moves into the alveoli. Carbon dioxide moves through the same path in the opposite direction and is exhaled.

## Asthma

Asthma is a disease that affects the tubes that bring air to and from the lungs, called airways. The airway tubes, the trachea, bronchi and bronchioles, are surrounded by muscles that can contract, narrowing the tubes and thereby making it difficult to breathe. An asthma attack is an acute narrowing of the tubes in response to exposure to an allergen (like pollen, dust, and animal dander), an irritant (like cigarette smoke, pollution, and cold air) or even just exercise. When the airways become inflamed, they can also swell and aggravate or sensitize the person to asthma. It is not clear as to what causes people to be prone to asthma, but heredity and environmental factors play a role. Asthma treatments seek to prevent or reverse the contraction of the tubes, reverse inflammation of the airways and can either provide long term alleviation or quick relief from an attack that is already occurring.

Indication	Company	Product	Phase
Asthma	<b>Aerovance Inc</b>	Aerovant / AER 001	II
	<b>APT Pharmaceuticals Inc</b>	ARD-1300 (hydroxychloroquine)	II
	<b>Aradigm Corp</b>	ARD-1300 (hydroxychloroquine)	II
	<b>Dynavax Technologies Corp</b>	Inhaled 1018 ISS	II
	<b>Genentech Inc</b>	HAE1 (PRO98498)	II
	<b>Gilead Sciences Inc</b>	Corus 1030 (lidocaine)	II
	<b>Inverseon Inc</b>	INV102	II
	<b>MAP Pharmaceuticals Inc</b>	MAP0010 / unit dose budesonide (UDB)	II
	<b>PDL BioPharma Inc</b>	Daclizumab	II
	<b>Theravance Inc</b>	GSK 597901	II
	<b>Theravance Inc</b>	GSK 678007	II
	<b>Theravance Inc</b>	GSK159797 / TD-3327	II
	<b>Theravance Inc</b>	GSK159802	II
	<b>Theravance Inc</b>	GSK642444	II
	<b>Genentech Inc</b>	Xolair (omalizumab)	III
	<b>MAP Pharmaceuticals Inc</b>	MAP0010 / unit dose budesonide (UDB)	III
	<b>DEY LP</b>	Zyflo CR (zileuton)	A
	<b>BioMarin Pharmaceutical Inc</b>	Orapred (prednisolone sodium phosphate)	M
	<b>BioMarin Pharmaceutical Inc</b>	Orapred ODT (prednisolone sodium phosphate)	M
	<b>DEY LP</b>	Cromolyn sodium Inhalation Solution	M
<b>DEY LP</b>	Metaproterenol Sulfate Inhalation Solution	M	
<b>DEY LP</b>	Zyflo (zileuton) tablet	M	
Respiratory Diseases (General)	<b>Novartis AG</b>	Streptopur	M

## Bronchospasm

Bronchospasm is characterized by difficulty in breathing due to the sudden constriction of the bronchi or bronchioles in the lungs. The constriction causes the airways to narrow and mucous production to increase, reducing the flow of air into the lungs and oxygen available to the body. This causes breathlessness, coughing and hypoxia. Bronchospasm usually accompanies asthma, chronic bronchitis, and anaphylaxis, and is a side effect of the drugs such as Pilocarpine or even aspirin.

Company	Product	Phase	Indication
DEY LP	AccuNeb (albuterol sulfate)	M	Bronchospasm
DEY LP	Albuterol Sulfate Inhalation Solution	M	
DEY LP	Ipratropium Bromide Inhalation Solution	M	
DEY LP	Metaproterenol Sulfate Inhalation Solution	M	
Impax Laboratories Inc	Terbutaline Sulfate Tablets	M	

## Chronic Obstructive Pulmonary Disease (COPD)

In normal breathing, the air sacs in the lungs at the end of the airways inflate upon inhalation and deflate again when a person exhales. A patient with Chronic Obstructive Pulmonary Disease is affected in four main ways: the air sacs are less elastic, so they do not deflate optimally; the walls between the air sacs break down, resulting in fewer, larger and thus less efficient air sacs; the airways become inflamed and swollen (chronic bronchitis); and more mucus is produced, which further obstructs the airways.

Company	Product	Phase	Indication
Aerovance Inc	Pulmolytic / AER 002	II	Chronic Obstructive Pulmonary Disease
Theravance Inc	GSK 597901	II	
Theravance Inc	GSK 678007	II	
Theravance Inc	GSK159797 / TD-3327	II	
Theravance Inc	GSK159802	II	
Theravance Inc	GSK642444	II	
Theravance Inc	GSK961081	II	
DEY LP	Perforomist Inhalation Solution (formoterol fumarate)	A	
DEY LP	DuoNeb (ipratropium bromide and albuterol sulfate)	M	

## Cough

Coughing is the body's way of removing foreign material or mucus from the airway tubes. Coughs have distinctive traits that can be recognized. A cough is only a symptom, not a disease, and often the importance of the cough can be determined only when other signs and symptoms are evaluated.

Company	Product	Phase	Indication
BIO-VED Pharmaceuticals Inc	Corshe	M	Cough

## Cystic Fibrosis

Cystic fibrosis is a hereditary disease that mainly affects the lungs and digestive system, causing progressive disability and for some, early death. Formerly known as cystic fibrosis of the pancreas, this entity has increasingly been labeled simply “cystic fibrosis.” Average life expectancy is around 37 years, although improvements in treatments mean a baby born today could expect to live longer.

Difficulty breathing and insufficient enzyme production in the pancreas are the most common symptoms. Thick mucus production as well as a less competent immune system results in frequent lung infections, which are treated, though not always cured, by oral and intravenous antibiotics and other medications. A multitude of other symptoms, including sinus infections, poor growth, diarrhea, and potential infertility (mostly in males, due to the condition Congenital bilateral absence of the vas deferens) result from the effects of CF on other parts of the body. Often, symptoms of CF appear in infancy and childhood and include meconium ileus, failure to thrive, and recurrent lung infections.

Cystic fibrosis is one of the most common life-shortening, childhood-onset inherited diseases. In the United States, 1 in 3900 children is born with CF. It is most common among Europeans and Ashkenazi Jews; one in twenty-two people of European descent carry one gene for CF, making it the most common genetic disease among such people.

Individuals with cystic fibrosis can be diagnosed prior to birth by genetic testing or in early childhood by a sweat test. Newborn screening tests are increasingly common and effective. There is no known cure for CF, and most individuals with cystic fibrosis die young — many in their 20s and 30s from lung failure, although with many new treatments being introduced the life expectancy of a person with CF is increasing. Ultimately, lung transplantation is often necessary as CF worsens.

CF is caused by a mutation in a gene called the cystic fibrosis transmembrane conductance regulator (CFTR). The product of this gene is a chloride ion channel important in creating sweat, digestive juices, and mucus. Although most people without CF have two working copies of the CFTR gene, only one is needed to prevent cystic fibrosis. CF develops when neither gene works normally. Therefore, CF is considered an autosomal recessive disease. The name cystic fibrosis refers to the characteristic ‘fibrosis’ (tissue scarring) and cyst formation within the pancreas, first recognized in the 1930s.

Indication	Company	Product	Phase
Cystic Fibrosis	<b>Aerovance Inc</b>	Aerolytic / AER 002	II
	<b>Gilead Sciences Inc</b>	Corus 1020 / Cayston (aztreonam lysine)	III
	<b>Nektar Therapeutics</b>	TMB100 / Tobramycin inhaled powder	III
	<b>Genentech Inc</b>	Pulmozyme (dornase alfa)	M
	<b>Novartis AG</b>	Tobi (tobramycin)	M
Mucolytic Agent	<b>DEY LP</b>	Sterile Acetylcysteine Solution	M

## Emphysema

Emphysema is a chronic lung disease most often caused by prolonged exposure to toxic chemicals and/or tobacco smoke. It is characterized by the loss of elasticity of the lung tissue, destruction of structures supporting the alveoli, the air sacs at the end of the airways, and destruction of blood capillaries feeding the alveoli. As a result, the small airways collapse during expiration and air gets trapped in the lungs. Symptoms include shortness of breath on exertion, hyperventilation, and an expanded chest.

Company	Product	Phase	Indication
Arriva Pharmaceuticals Inc	Recombinant alpha 1-antitrypsin / rAAT	II	Emphysema

## Pulmonary Embolism

Pulmonary embolism is the blockage of a pulmonary artery in the lungs by a blood clot (thrombus), fat, air, or tumor cells. The most common form is a blood clot which originated as a deep vein thrombus and traveled through the veins and right heart into the lungs. Most patients are diagnosed through pulmonary angiopathy and CT scans. Symptoms include difficulty breathing, pain during breathing, and circulatory instability.

Company	Product	Phase	Indication
Genentech Inc	Activase (alteplase)	M	Pulmonary Embolism

# INTERMUNE<sup>®</sup>

On behalf of InterMune, Inc. (NASDAQ: ITMN) we are pleased to participate with Bay Bio in the important effort to build a greater understanding of the biotechnology industry and the ways we're working to create change in patients' lives.

InterMune, Inc. is dedicated to developing and commercializing innovative therapies in pulmonology and hepatology. Pulmonology is the field of medicine concerned with the diagnosis and treatment of lung conditions, while hepatology is concerned with the diagnosis and treatment of disorders of the liver.

Our pulmonology pipeline includes the fully enrolled Phase 3 "CAPACITY" program, which is evaluating our product candidate pirfenidone for the treatment of patients with idiopathic pulmonary fibrosis, or IPF. IPF is a chronic, fatal lung disease characterized by inflammation and scarring of the lungs, for which no treatments are currently approved, either in the United States or Europe. InterMune also has an exciting research program focused on small molecules for pulmonary disease.

In hepatology, InterMune's development portfolio includes the hepatitis C virus (HCV) protease inhibitor compound ITMN-191, which has completed a Phase 1a clinical trial in healthy subjects. We are excited to be among the leaders in developing protease inhibitors, a new class of compounds that has the potential to fundamentally change the treatment paradigm and improve the rate of success in curing patients chronically infected with the hepatitis C virus. ITMN-191 is being developed in collaboration with our partner, Roche. InterMune also has a second-generation HCV protease inhibitor research program and a research program evaluating a new target in hepatology.

Learn more about our mission, our product candidates and our people at [www.intermune.com](http://www.intermune.com).

## Pulmonary Fibrosis

Pulmonary fibrosis is an inflammatory lung disorder characterized by abnormal formation of fibrous tissue (fibrosis) between the tiny air sacs (alveoli) or ducts of the lungs, affecting the lungs' ability to move oxygen into the bloodstream. Currently in the United States there are over 200,000 patients with pulmonary fibrosis, though the number may be much higher due to misdiagnosis.

Indication	Company	Product	Phase
Pulmonary Fibrosis	<b>InterMune Inc</b>	S-7701 (pirfenidone)	III

## Pulmonary Hypertension

Pulmonary hypertension is an increase in blood pressure in the lungs's main artery. It can be a very severe disease marked by decreased exercise tolerance and right-sided heart failure. Symptoms include shortness of breath, fatigue, cough, angina, fainting, and edema. The most common cause of pulmonary hypertension is left heart failure due to a malfunction of the left ventricle, though ischemic heart disease, scleroderma, and other autoimmune disorders can also lead to this disease. Pulmonary embolism can also be a factor.

Indication	Company	Product	Phase
Pulmonary Hypertension	<b>Gilead Sciences Inc</b>	Ambrisentan	PA
	<b>Gilead Sciences Inc</b>	Volibris (ambrisentan)	PA
	<b>Gilead Sciences Inc</b>	Letairis / Volibris (ambrisentan)	A
	<b>Gilead Sciences Inc</b>	Flolan (epoprostenol sodium)	M
	<b>Genentech Inc</b>	Tracleer (bosentan)	PM

## Respiratory Distress Syndrome

Respiratory Distress Syndrome is a serious reaction to injuries of the lung and results in increased permeability and pulmonary edema. It is characterized by inflammation of the lung leading to impaired gas exchange and multiple organ failure. The condition is life threatening and often lethal, requiring mechanical ventilation.

Indication	Company	Product	Phase
Respiratory Distress Syndrome	<b>Genentech Inc</b>	TNX-832	II
	<b>DEY LP</b>	Curosurf (poractant alfa) Intratracheal Suspension	M