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# PRIMARY CILIARY DYSKINESIA/KATAGENERS DISEASE

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• **ABSTRACT :**

Certainly! This article talks about a rare condition called Primary Ciliary Dyskinesia (PCD). It's a disease where the tiny hair-like structures in our airways don't work properly. This can lead to problems with breathing, infections, and other issues.

PCD is quite rare, affecting about 1 in 10,000 to 1 in 20,000 kids, maybe even more. It can be tricky to diagnose because there's no one perfect test for it. Doctors often have to use a combination of tests and look at genetic factors too.

One interesting thing mentioned is that scientists are exploring a way to fix the genes that cause PCD, which could be a game-changer for treatment in the future.

Right now, there's no specific treatment proven by big scientific trials, so doctors usually treat it like they do with some other lung diseases. They focus on keeping the airways clear, using things like saltwater sprays and chest therapy. Plus, they use antibiotics when needed.

The article also highlights how important it is for doctors from different specialties to work together and for patients to go to specialized centers for the best care. There's a hope that by improving how we diagnose and treat PCD, we can make life better for those who have it.

**Keywords :**

Cytofila,Kartagener syndrome, Left-right asymmetry, Nitric oxide

• **INTRODUCTION :**

• Primary Ciliary Dyskinesia (PCD): A rare, inherited genetic disorder where the cilia (tiny hair-like structures) in the lungs, ears, and sinuses do not move properly. This leads to mucus buildup, increasing the risk of infections.

• Kartagener's Syndrome: A subset of PCD. People with this syndrome have the triad of chronic sinusitis, bronchiectasis, and situs inversus (which means their internal organs are reversed or mirrored from their normal positions).Kartagener's syndrome (KS) is a

subset of a larger group of ciliary motility disorders called primary ciliary dyskinesias(PCDs).

- Bronchiectasis: A long-term condition where the airways in the lungs become widened and lose their elasticity, causing mucus to build up. This can lead to chronic coughing, breathlessness, and repeated lung infections.
- Ciliopathy: A broader term for disorders caused by defects in the function or structure of cilia. PCD is a type of ciliopathy.
- Mucociliary Clearance: The process by which mucus and trapped inhaled particles are transported out of the lungs. Cilia beat in coordinated waves to move mucus up and out of the respiratory tract. In condition like PCD, this process is disrupted, leading to mucus accumulation and a higher risk of respiratory infections.
- Left-right asymmetry (LR asymmetry) is the process in early embryonic development that breaks the normal symmetry in the bilateral embryo. In vertebrates, left-right asymmetry is established early in development at a structure called the left-right organizer (the name of which varies between species) and leads to activation of different signalling pathways on the left and right of the embryo.
- Primary Ciliary Dyskinesia (PCD):

PCD is a condition where tiny hair-like structures, called cilia, that move mucus around inside our bodies, don't work properly. This can cause problems in the lungs, ears, and sinuses, and can also lead to fertility issues.

How did we learn about PCD?

- A doctor named Kartagener noticed some patients had sinus problems, lung issues, and organs on the opposite side of their body.
- Later, another doctor, Afzelius, found out that the issues were because of faulty cilia.

Why do people get PCD?

Most of the time, it's because of genes passed down from parents. Sometimes, it's because of certain gene changes on the X-chromosome.

**How do doctors find out if someone has PCD?**

- By looking at symptoms: like constant coughs, sinus problems, or issues having children.
- By checking how cilia move under a microscope.
- By looking at a person's genes.

How is PCD treated?

- By helping to clear mucus from the lungs.
- Using antibiotics for infections.
- Regular doctor visits to check lung health.

What's next for PCD?

Scientists are looking for better ways to treat PCD, maybe even by fixing the faulty genes.

PCD is a condition where the body's cilia don't work well, leading to health problems. It's important for doctors to know about it so they can help patients get better

## Cilia Biology:

### Structure and Function :

Primary Ciliary Dyskinesia (PCD) is a rare genetic disorder where the tiny hair-like structures called cilia, which normally help move mucus and clear the airways,don't work properly.
People with PCD might experience chronic respiratory problems, fertility issues, and sometimes unusual positioning of internal organs. It's not easy to diagnose, and there'sno specific treatment proven by clinical trials yet.
cus

Researchers are making progress in understanding the genes linked to PCD, and there's hope for future treatments, like gene editing, to improve cilia function. The goal is to raise awareness among doctors so that they can recognize and manage PCD early, potentially preventing severe lung damage.

**Epidemiology** :

In 2010, a big study looked at kids with a lung disease called PCD in 26 European countries. They found that the number of kids diagnosed varied a lot, from 1 in 10,000 to 1 in 20,000 born. The overall estimates for the disease ranged widely, from1 in 2,200 to 1 in 40,000, because different ways of studying it gave different results.

Cyprus, Switzerland, and Denmark had the most kids with PCD. The differences in diagnosis could be because of where people live, specific gene issues, marriages between close relatives, or how each country checks for PCD.

PCD is a condition where tiny hair-like structures in the body don't work properly. Getting diagnosed can be a problem because many doctors might not be aware of it.

Europe, kids with this condition are usually diagnosed around 5 years old, but it takes longer if they also have a condition called situs inversus (SI). Sometimes, people have to see a lot of doctors (50-100 visits) before finding out they have PCD. Even in specializedcenters, awareness of this disorder seems to be an issue.

Creating a list of information about a rare condition called Primary Ciliary Dyskinesia (PCD) is really important. This list, or registry, started in January 2014 and includes details about 201 PCD patients from various places in Europe and North America. This helps gather data on how common PCD is, how it shows up in patients, and what kinds of treatments are being used.

**Disease Manifestations ;**

Sure, I can simplify it for you. Primary Ciliary Dyskinesia (PCD) is a rare genetic disorder affecting cilia, tiny hair-like structures in our body. These cilia usually help in moving mu and other substances in our airways. In PCD, these cilia don't work properly, leading to respiratory issues, infections, and other complications.

Diagnosing PCD can be challenging, involving clinical observations, genetic analysis, and specialized tests. Treatment focuses on managing symptoms, and there's ongoing research exploring potential gene-editing approached

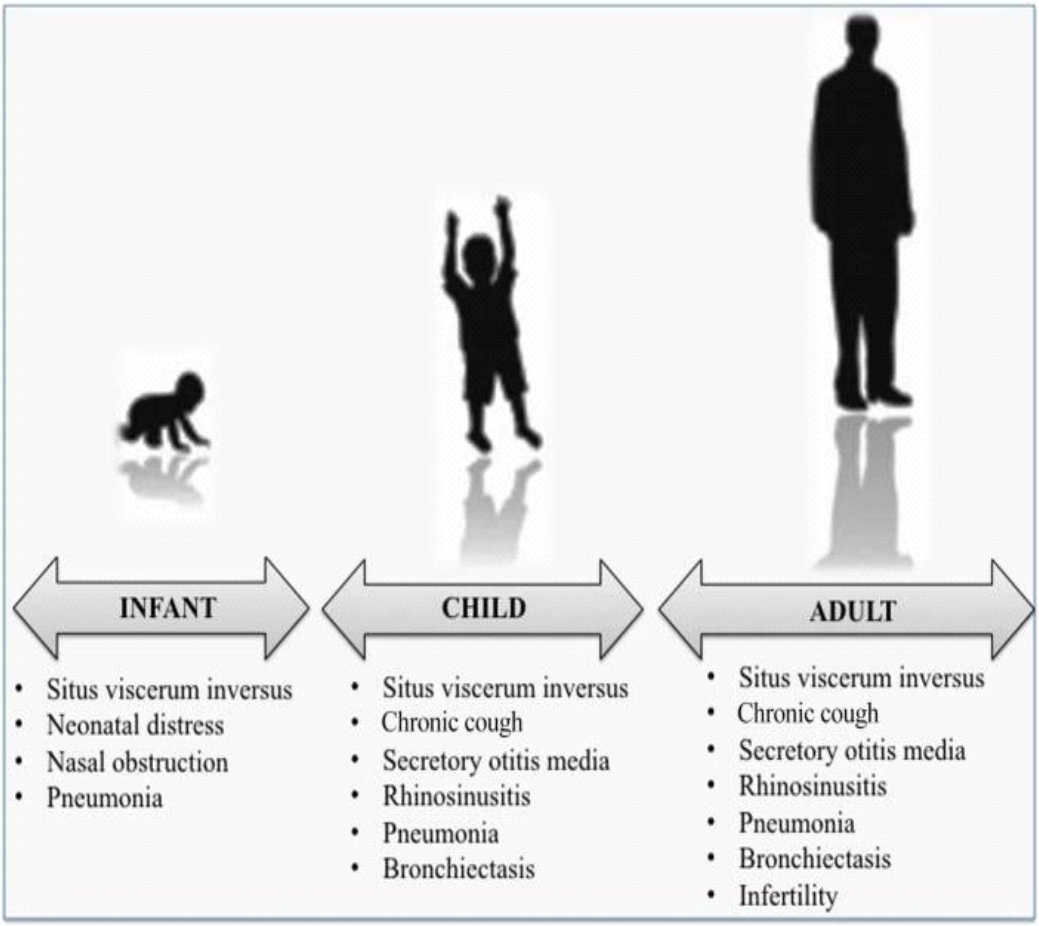



Figure no 1 : Model

Newborns with PCD might have breathing problems that last longer compared to common respiratory issues in babies. Unlike usual cases where difficulties breathing after birth resolve by the fifth day, PCD-related challenges can be more severe and mayinvolve pneumonia and lobar collapse, requiring a longer hospital stay.

Kids with PCD often have a runny nose for a long time, which can cause them to lose their sense of smell. They also tend to get ear infections a lot, and some may need antibiotics more than 30 times in their life. These issues can lead to problems like trouble breathing during sleep.

Lower airways are commonly involved in PCD.

Kids with PCD often cough a lot because they get bronchitis or pneumonia frequently. This can lead to a lasting lung issue called bronchiectasis, where their airways are damaged. Doctors use scans to check their lungs, but some tests involve more radiation. Kids with PCD might also have asthma that's tough to treat, and tests can show blockages in their airways.

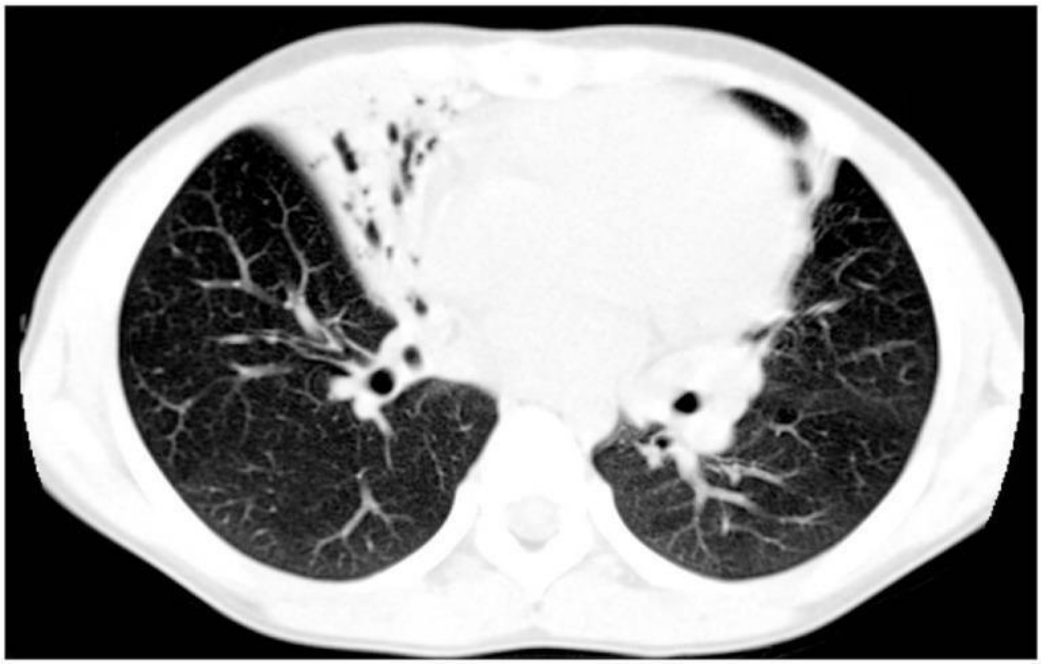


Figure 2 : High-resolution computed tomography findings from a 7-year-old girl with primary ciliarydyskinesia.

The scan demonstrates an area of consolidation both in the lingula and in the middle lobe, the latter also including bronchiectasis.

When checking the mucus from the lungs of kids with PCD, it often has certain bacteria when they are young, like Haemophilus influenzae and Streptococcus pneumoniae. As they grow up, a different bacteria called Pseudomonas aeruginosa becomes more common, and in some cases, it gets sticky. This doesn't happen a lot, but about 5% of PCD patients under

19 have the sticky kind. Also, although it's rare in childhood, more than 10% of adults with PCD can have another type of bacteria called non-tuberculous mycobacteria in their mucus.

Some guys with PCD may find it hard to have babies because their sperm doesn't work properly. The tiny hair-like part of the sperm, called the tail, might not move well due to issues with cilia. Common problems include missing parts and changes in the tubes. Guys with PCD often have sperm that doesn't move a lot, making it difficult to have a baby naturally. So, if they want to become parents, they might need help from procedures like artificial insemination or in vitro fertilization. It's important for couples to get advice and check the guy's sperm before trying these methods. Women with PCD may have a higher chance of ectopic pregnancies and reduced ability to get pregnant because of issues withcilia in the egg pathway.

Primary Ciliary Dyskinesia (PCD), where hair-like structures in the body don't work properly. It can cause breathing and lung problems, fertility issues, and other complications. Doctors diagnose it using a mix of clinical information and special tests. The text also mentions the need for doctors to be aware of the condition to catch it early and manage it better.

To keep tabs on lung conditions like PCD, we usually use spirometry, a common test.
Recently, there's interest in the lung clearance index (LCI), which is sensitive to early airway issues. Yet, the data on LCI, spirometry, and lung structure changes in PCD are a bit confusing. We need more research to understand how LCI plays a role in the disease's progression over time.

## Diagnosis :

It's crucial to conduct a comprehensive diagnostic evaluation for Primary Ciliary Dyskinesia (PCD), especially when there's a positive family history, as familial cases can contribute to 10% of PCD diagnoses. Siblings of affected individuals should undergo assessment, especially if they show subtle respiratory symptoms that might not immediatelysuggest PCD.

There's a questionnaire called PICADAR that helps predict if someone with a persistent wet cough might have Primary Ciliary Dyskinesia (PCD). It looks at things like birth history,
respiratory symptoms, and more. If the score is 10 or higher, there's a high chance (over 90%) of having PCD, and a score of 5 or higher suggests more than an 11% chance of havingPCD.

Finding out if someone has Primary Ciliary Dyskinesia (PCD) isn't easy because there isn't a single test that can confirm it. Doctors use a mix of complicated tests like checking nasal nitric oxide, high-speed video microscopy, and transmission electron microscopy to piece together the diagnosis.

Tests like the saccharine test and using a radioactive tracer were once common for diagnosing Primary Ciliary Dyskinesia (PCD). They aimed to show problems with how mucus moves due to issues with cilia motion. However, the saccharin test might miss some cases, and the radioactive tracer test exposes patients to a bit of radiation. Because of limited
evidence, doctors don't strongly recommend these tests for PCD anymore.

In the past, to diagnose Primary Ciliary Dyskinesia (PCD), doctors used to analyze tiny sections of cilia with a transmission electron microscope. They usually took samples from the nose or lower respiratory tract using a brush or during bronchoscopy. This detailed analysis helped identify specific defects in cilia structure.

However, this method, while powerful, couldn't catch all PCD variants (around 30% were missed), and some abnormalities could be caused by infections or inflammation. If the initial test shows normal results but there's a strong suspicion of PCD based on symptoms, furthertests are recommended.

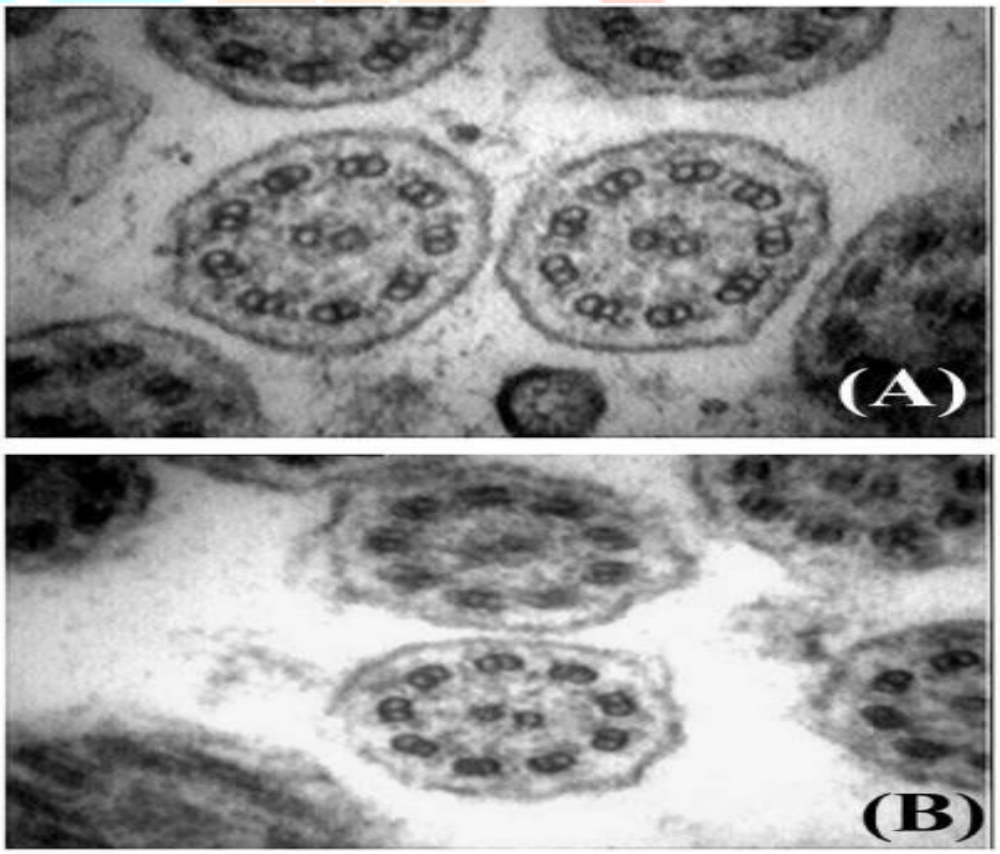



Figure 3 .Electron microscopy findings showing normal cilia ultrastructure from an healthy subject (A), and outer and inner dynein arms defect from a patient with primary ciliary dyskinesia (B) (courtesy of Dr. Mariarosaria Cervasio, Department of Advanced Biomedical Sciences, Anatomo-Pathology Unit, Federico II University, Naples, Italy).

To accurately diagnose Primary Ciliary Dyskinesia (PCD) using High-Speed Video Microscopy Analysis (HVMA), doctors suggest repeating the assessment after air–liquid interface culture. It's important not to rely solely on Ciliary Beat Frequency (CBF) but also evaluate Ciliary Beat Pattern (CBP) for a complete diagnosis.

Different medical centers use various protocols for HVMA, involving different tools and criteria. Generally, a CBF below 11 beats per second may indicate an issue, but HVMA aloneisn't enough to confirm or rule out PCD.

Scientists are now using a new method called high-resolution immunofluorescence (IF) analysis to study where certain proteins are in respiratory cells. This method is good at finding structural issues, and it can even detect problems with specific proteins that other methods might miss. More labs are starting to use this technology, and as it continues to improve, it may help identify even more variations of Primary Ciliary Dyskinesia (PCD).

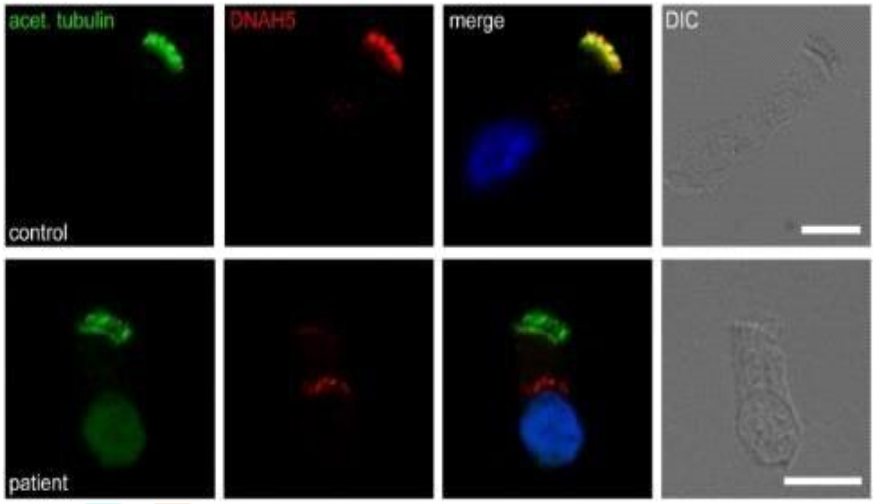



Figure 4.Immunofluorescence staining of human respiratory epithelial cells with DNAH5-specific antibodies (red) and antibodies against acetylated α-tubulin (green). Nuclei were stained with Hoechst 33342 (blue). Overlays and bright-field images are shown on the right.

Whereas in healthy human respiratory epithelial cells (control; upper panel), both antibodies colocalize along the entire length of the ciliary axonemes, in an individual with an outer dynein arm defect (patient; lower panel), DNAH5 is absent.

**Genetics :**

PCD, a genetic condition, often comes from both parents passing on a certain gene issue. There are many genes linked to it, and some mutations make the condition more likely. Figuring out which mutations cause the problem can be tough. Most mutations are unique to each person, and they don't often gather in specific areas.

Certain mutations match up with what we see under microscopes. Some mutations make the condition less severe, like those in the RSPH1 gene. Interestingly, some mutations don't mess up how the body is shaped. For example, some don't affect sperm movement, making affected men fertile. If there are issues with multiple cilia, it usually means more serious breathing problems.

Research is ongoing, and we're learning that lung problems can vary, with some mutations making things worse than others.

Table 1 :

Genes associated with primary ciliary dyskinesia and corresponding ultrastructure.

| Genea. | Axonemal/cellular structure or function |
|---|---|
| DNAH5, DNAI1, DNAI2, DNAL1, NME8(TXNDC3) | Outer dynein arm (ODA) subunit |
| CCDC114, ARMC4, CCDC151, TTC25 | ODA targeting/docking factor |
| DNAAF1 (LRRC50), DNAAF2 (KTU), DNAAF3, HEATR2, LRRC6, ZMYND10, DYX1C1 (DNAAF4), SPAG1, CCDC103, C21ORF59 | Cytoplasmic dynein arm assembly ortransport factor |
| RSPH1, RSPH3, RSPH4A, RSPH9 | RSPH subunit |
| CCDC39, CCDC40 | NL/DRC factor |
| CCDC164, CCDC65 | NL subunit |

| | |
|---|---|
| DNAH11 | ODA subunit |
| HYDIN | CP subunit |
| CCNO, MCIDAS | CCNO: cytoplasmic centriole assembly and docking factor; MCIDAS: nuclear regulator of CCNO and FOXJ1 |
| OFD1, RPGR | Functions related to non-motile cilia; role in motile cilia unknown |

Modern genetic tools can find the cause of PCD in around 70% of affected people. Using a method called next-generation sequencing, which is cost-effective, makes diagnosing PCD easier than doing many different tests. As technology improves, we can expect to get evenbetter at finding PCD early and treating it sooner.

**Current and Future Treatment Strategies :**

Treating Primary Ciliary Dyskinesia (PCD) is tricky because there aren't specific guidelines for it. Doctors often rely on expert opinions or use evidence from Cystic Fibrosis (CF), even though PCD and CF have different underlying issues. Having specific recommendations for PCD would help in better, more personalized treatments.

Treating Primary Ciliary Dyskinesia (PCD) involves keeping airways clear, preventing infections, and avoiding triggers like secondhand smoke. Different methods, such as chest physiotherapy, postural drainage, and breathing exercises, are used to help clear the airwayseffectively.

Physiotherapists use different methods like postural drainage, percussion, and breathing exercises to help clear mucus in Primary Ciliary Dyskinesia (PCD). There are devices like Positive Expiratory Pressure (PEP) valves and oscillating vests that can assist patients in getting rid of mucus. Despite not having clear evidence on which method is best, it's recommended for PCD patients to do daily physiotherapy.

People with obstructive pulmonary disease (OPD) should exercise regularly to make their breathing muscles stronger and keep their lungs healthy. Many OPD cases face difficulties in doing strenuous activities, and about half of them spend less than 3 hours per week on physical activity, indicating low levels of exercise. Engaging in exercise before airway clearance can be more effective in improving mucus clearance and acting as a bronchodilator compared to certain medications. It's important to address the lower oxygen uptake observed in OPD patients, highlighting the need for tailored exercise programs to improve their overall respiratory function.

Inhaling a mist through a nebulizer is a common way to moisten and thin thick mucus in the airways, helping with clearing it out. For people with bronchiectasis, breathing in salty mist (hypertonic saline) can make it easier to clear mucus. In a study, adults with bronchiectasis using hypertonic saline reported improved mucus clearance, used fewer antibiotics, and had fewer emergency visits. However, it didn't show big changes in breathing tests, mucus bacteria, or overall quality of life. A similar study with salty mist in PCD patients didn't improve their quality of life or significantly change breathing tests or airway inflammation. We need more and larger studies, especially including children, to be sure about these results.

For those with Primary Ciliary Dyskinesia (PCD), it's crucial to prevent infections, just like with other ongoing lung issues. PCD patients, both kids and adults, are more at risk for certain infections, so they should get specific vaccines to protect against them. This includes vaccinations for pneumococcal disease and getting the flu shot every year. Following the routine vaccination schedule for your area is also important. These steps help lower the chances of respiratory infections in people with PCD.

Taking care of children and adults with Primary Ciliary Dyskinesia (PCD) works best when a team of different experts works together. This team could include doctors who specialize in kids (pediatricians), lung doctors (pulmonologists), therapists for the chest (chest physiotherapists), genetics experts (geneticists), scientists (biologists), heart doctors (cardiologists), imaging experts (radiologists), men's health doctors (andrologists), and ear, nose, and throat surgeons

(ENT surgeons). By combining their skills, these specialists can work together to give the best care for people with PCD.

**Psychological Issues:**

Doctors taking care of kids with PCD need to understand that the disease not only affects the child's health but also has a big impact on how the whole family lives. PCD leads to breathing problems that get worse over time, and this can be tough for both the child and their family. Especially during teenage and young adult years, dealing with a long-term condition like PCD can be emotionally challenging. The good news is, if PCD is diagnosed early and gets the right treatment, it usually helps improve the overall outcome for the patient.

**Ethics Statement :**

The authors made sure to get permission in writing from the patients, healthy individuals, or their legal guardians before publishing any images.

**Harbal APPROACHES :**

**1) Echinacea :**

Echinacea, or coneflower, is a flowering plant and popular herbal remedy

Scientific name : Echinacea purpurea

Parts used : leaf, flower, and root

Important comments :

MOA :

1)Phagocytosis activation, fibroblast stimulation and increased respiratory activity all contribute to increased leukocyte motility.

2)boost immune function, relieve pain, reduce inflammation, and have hormonal, antiviral, and antioxidant effects.

**2) GINSENG :**

**GINSENG IS A MEDICINAL PLANT WHOSE ROOTS ARE USUALLY STEEPED TO MAKE A TEA OR DRIED TO MAKE A POWDER.**

Scientific name ; Ashwagandha Indian ginseng Parts used : roots and rhizomes,


**MOA:**

1)Y-amino butyric acid agonist there by having sedative effects on CNS

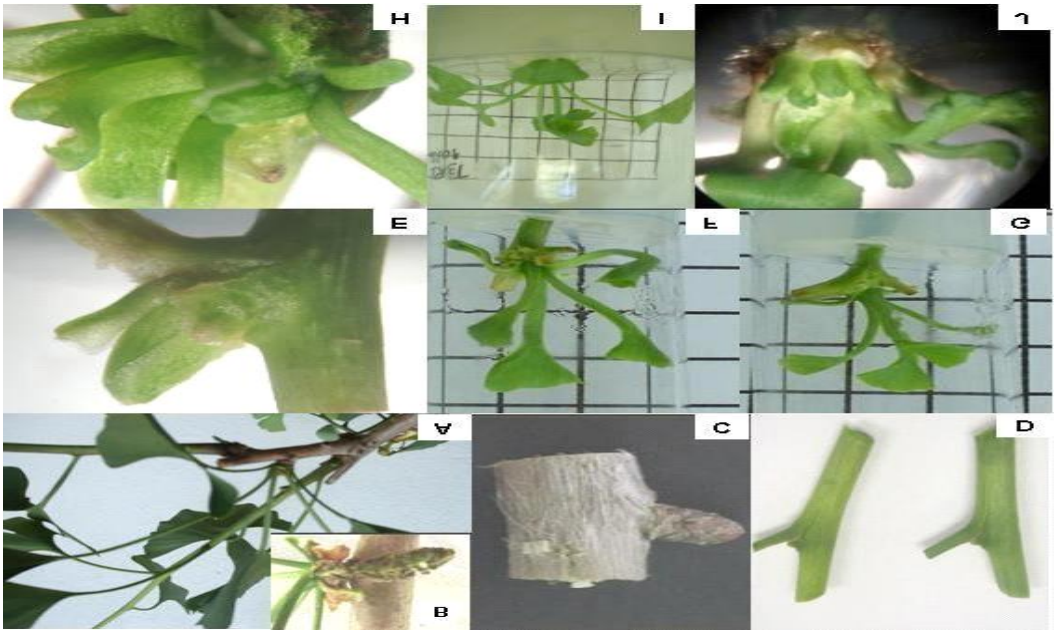
2) ashwagandha may have an attenuating effect on hypothalamic pituitary(HPA) adrenal axis activity in response to a stresser the HPA axis is associated with a series of response ultimately leading to increase in both cortisol and DHEA [31,32] concentrations

**3) GINKGO BILOBA :**

BILOBA, ALSO KNOWN SIMPLY AS GINKGO, IS AN HERBAL MEDICINE DERIVED FROM THE MAIDENHAIR TREE (8TRUSTED SOURCE).

Scientific name ; ginkgo,gingko

Parts used : extract prepare from its fan-shaped leaves Important compound : quercetin,kaempferol,isorhamnetin



**MOA :**

- Acts as a free radical scavenger, protecting neurones from oxidative damage and apoptosis related to aging, cerebral ischemic and neurodegenerative disorders.
- Gingko biloba also inhibit amyloid -B neurotoxicity and protects against hypoxic challenges and increase oxidative stress

**4) ELDERBERRY** :

Elderberry is an ancient herbal medicine typically made from the cooked fruit of the Sambucus nigra plant. It has long been used to relieve headaches, nerve pain, toothaches, colds, viral infections, and constipation (10).

PARTSCIENTIFIC S USED : FLOWERS
IMPORTANT CONAME ; SAMBUCUS IS A GENUS OF FLOWERING PLANTS IN THE FAMILY ADOXACEAE.
MPOUND : THE MAIN POLYPHENOLS IN ELDERBERRY FRUIT ARE CHLOROGENIC ACID, NEOCHLOROGENIC ACID,
CRYPTOCHLOROGENIC ACID, QUERCETIN, QUERCETIN-3-
RUTINOSIDE (RUTIN), QUERCETIN-3-GLUCOSIDE (ISOQUERCITRIN), KAEMPFEROL-3-RUTINOSIDE, KAEMPFEROL-3-GLUCOSIDE (ASTRAGALINE), ISORHAMNETIN-3-RUTINOSIDE AND
ISORHAMNETIN-3-GLUCOSIDE.

**MOA**:

1) ELDERBERRY INHIBITS H1N1 ACTIVITIES BY BINDING TO H1N1 VIRIONS AS WELL AS BY BLOCKING HOST CELL RECOGNITION ANDENTRY (5).
2) IT MAY ALSO PREVENT HIV1 INFECTION BY BINDING TO VIRAL GLYCOPROTEINS SUCH AS GP120 (10), BUT ADDITIONAL INVESTIGATIONS ARE REQUIRED TO CLARIFY THOSE MECHANISMS

**5) St. John's wort :**

St. John's wort (SJW) is an herbal medicine derived from the flowering plant Hypericum perforatum. Its small, yellow flowers are commonly used to make teas, capsules,or extracts (16Trusted Source).

SCIENTIFIC NAME ; Hypericum perforatumPARTS USED : stem, petals, and flowers.

IMPORTANT COMPOUND : hypericin, hyperforin, and various flavonoids such as quercetin,rutin, and kaempferol.

**MOA:**

1) John's Wort acts as a reuptake inhibitor of serotonin, dopamine, and norepinephrine.

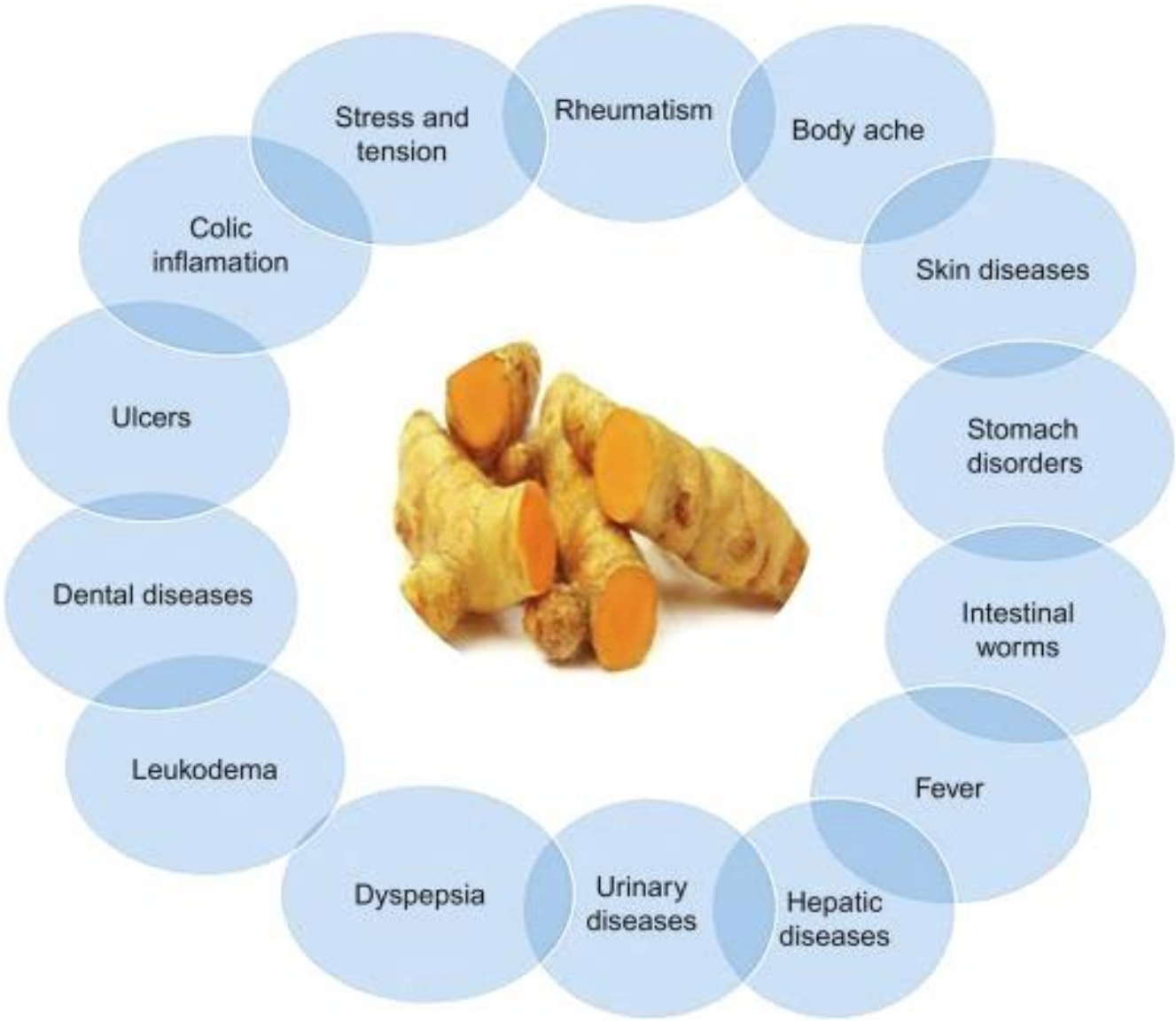
2) Elevated levels of neurotransmitters are believed to be helpful when treatingdepression.

3) neuronal synapses, as well as dopamine and norepinephrine.

**6) Turmeric :**

Turmeric (Curcuma longa) is an herb that belongs to the ginger family(18Trusted Source).

SCIENTIFIC NAME : Curcuma longa and Curcuma aromatica
PARTS USED : finger-like underground rhizomes are dried
IMPORTANT COMPOUND : Curcumin, a yellow polyphenolic pigment from the Curcuma longa L. (turmeric) rhizome, has been used for centuries for culinary and food coloring purposes, and as an ingredient for various medicinal preparations, widely used in Ayurvedaand Chinese medicine.



**MOA:**

- colorful plant-based foods, turmeric is rich in phytonutrients that may protect the body.
- neutralizing free radicals (pollution, sunlight) and shielding the cells from damage."
- Diets rich in plant-based foods are associated with prevention of medical conditions such as cancer and heart disease.

**7) Ginger:**

Ginger is a commonplace ingredient and herbal medicine. You can eat it fresh or dried, though its main medicinal forms are as a tea or capsule.

SCIENTIFIC NAME : Zingiber officinale

PARTS USED : The ginger spice comes from the roots of the plant. It's used as a foodflavoring and medicine.

Rhizome, ginger root or ginger

IMPORTANT COMPOUND : Eating ginger can cut down on fermentation, constipation and other causes of bloating and intestinal gas.

Phenolic and terpene compounds.

**MOA:**

1)Ginger and its metabolites appear to accumulate in the gastrointestinal tract and exert their effects.

2)relieving pain through anti-inflammatory effects, soothing the digestive system through carminative effects, and alleviating nausea

.

**~~8) Valerian :~~**

Sometimes referred to as "nature's Valium," valerian is a flowering plant whose roots are thought to induce tranquility and a sense of calm.

SCIENTIFIC NAME : Valeriana officinalis

PARTS USED :roots and rhizomes (underground stems)

IMPORTANT COMPOUND : essential oils, iridoid compounds such as monoterpenes and sesquiterpenes, flavonoids, alkaloids, amino acids, and lignanoids.

Valerian Flowers

**MOA :**

- Valerian is a medicinal herb that produces anxiolytic and sedative effects.
- It was suggested that valerian acts via gamma-aminobutyric acid (GABA)ergicmechanisms.
- Previous studies showed binding of valerian extract to GABA receptors, but the functional effect of the binding has not been demonstrated.

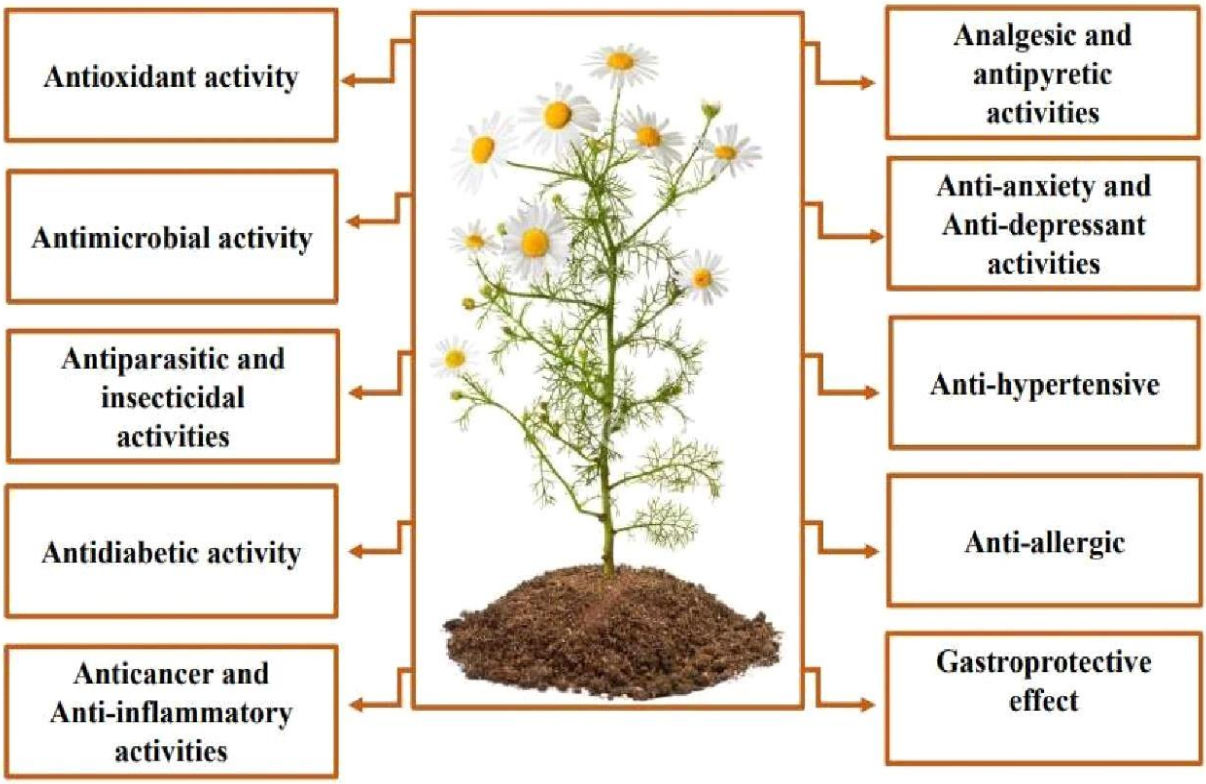
**9) Chamomile :**

Chamomile is a flowering plant that also happens to be one of the most popularherbal medicines in the world.

SCIENTIFIC NAME :genus Chamomilla

PARTS USED :the white and yellow flower head.

IMPORTANT COMPOUND : quercetin, apigenin, luteolin, and rutin.




**MOA :**

Chamomile flowers are abundant in chamazulen and α-bisabolol,

antibacterial, anti-inflammatory, wound healing, and mood-regulating properties.

**Acknowledgments :**

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Author Contributions :

VM helped plan and design, wrote part of the manuscript, and gave the OK for publishing. CW wrote some of the manuscript and approved it for publication. FS played a major role in planning, drafting, and revising the manuscript, and approved the final version for publishing.

Conflict of Interest Statement :

The research was carried out without any involvement or connections to businesses or financial interests that could create a conflict of interest.

**Conclusion**

Primary Ciliary Dyskinesia (PCD) is a rare genetic disorder affecting 1 in 10,000 to 1 in 20,000 children. It disrupts mucociliary clearance, leading to mucus accumulation and increased infection risk. The condition was discovered by Kartagener and Afzelius due to faulty cilia. Diagnosis involves observing symptoms and examining cilia movement. Treatment involves clearing mucus from the lungs. There is no specific treatment yet, but researchers are working on understanding the genes linked to PCD and hope for future treatments like gene editing. The goal is to raise awareness among doctors to recognize and manage PCD early.Primary Ciliary Dyskinesia (PCD) is a condition where hair-like structures in the body don't work properly, causing breathing and lung problems, fertility issues, and other complications. It is diagnosed using a mix of clinical information and special tests, with spirometry being a common test. The lung clearance index (LCI) is sensitive to early airway issues, but data on LCI, spirometry, and lung structure changes in PCD are confusing.Diagnosis is crucial, especially when there's a positive family history, as familial cases can contribute to 10% of PCD diagnoses. A questionnaire called PICADAR helps predict if someone with a persistent wet cough might have PCD. Finding out if someone has PCD is difficult due to the lack of a single test that can confirm it. Doctors use a mix of complicated tests like checking nasal nitric oxide, high-speed video microscopy, and transmission electron microscopy to piece together the diagnosis.Tests like the saccharine test and using a radioactive tracer were once common for diagnosing PCD, but they might miss some cases and expose patients to radiation. In the past, doctors used to analyze tiny sections of cilia with a transmission electron microscope, but this method couldn't catch all PCD variants and some abnormalities could be caused by infections or inflammation.

**References :**


- This is a citation for a document about primary ciliary dyskinesia (PCD), providing recommendations for diagnosis, monitoring, and treatment. The publication is from 2016 and was published in the Pediatric Pulmonology journal.

- This citation is from a study about diagnosing primary ciliary dyskinesia (PCD) using advanced genetic sequencing. The research looked at both the genetic and clinical findings in Italian patients. The study was published in the Journal of Molecular Diagnostics in 2016.

- Moore A, Escudier E, Roger G, Tamalet A, Pelosse B, Marlin S, et al. RPGR is mutated in patients with a complex X linked phenotype combining primary ciliary dyskinesia and retinitis pigmentosa. J Med Genet (2006) 43:326–33. 10.1136/jmg.2005.034868[PMC free article] [PubMed] [CrossRef] [Google Scholar]

- Paff T, Loges NT, Aprea I, Wu K, Bakey Z, Haarman EG, et al. Mutations in PIH1D3 cause X-linked primary ciliary dyskinesia with outer and inner dynein arm defects.Am J Hum Genet (2017) 100:160–8.

- Kartagener M. Zur pathogenese der bronkiectasien. Bronkiectasien bei situs viscerum inversus. Beitr Klin Tuberk Spezif Tuberkuloseforsch (1933) 83:489–501.

- Afzelius BA. A human syndrome caused by immotile cilia. Science (1976) 193:317–9. 10.1126/science.1084576 [PubMed] [CrossRef] [Google Scholar]

- Eliasson R, Mossberg B, Camner P, Afzelius BA. The immotile-cilia syndrome. A congenital ciliary abnormality as an etiologic factor in chronic airway infections and male sterility. N Engl J Med (1977) 297:1–6. 10.1056/NEJM197707072970101 [PubMed] [CrossRef] [Google Scholar]

- Zimmerman K, Yoder BK. SnapShot: sensing and signaling by cilia. Cell (2015) 161:692. 10.1016/j.cell.2015.04.015 [PMC free article] [PubMed] [CrossRef] [Google Scholar]

- Mitchison HM, Valente EM. Motile and non-motile cilia in human pathology: from function to phenotypes. J Pathol (2017) 241:294–309. 10.1002/path.4843 [PubMed][CrossRef] [Google Scholar]

- Raidt J, Wallmeier J, Hjeij R, Onnebrink JG, Pennekamp P, Loges NT, et al. Ciliary beat pattern and frequency in genetic variants of primary ciliary dyskinesia. Eur Respir J (2014) 44:1579–88. 10.1183/09031936.00052014 [PubMed] [CrossRef] [Google Scholar]

- Horani A, Ferkol TW, Dutcher SK, Brody SL. Genetics and biology of primary ciliary dyskinesia. Paediatr Respir Rev (2016) 18:18–24. 10.1016/j.prrv.2015.09.001 [PMC freearticle] [PubMed] [CrossRef] [Google Scholar]

- Ferkol TW, Puffenberger EG, Lie H, Helms C, Strauss KA, Bowcock A, et al. Primary ciliary dyskinesia-causing mutations in Amish and Mennonite communities. J Pediatr (2013) 163:383–7. 10.1016/j.jpeds.2013.01.061 [PMC free article] [PubMed] [CrossRef] [Google Scholar]

- Kennedy MP, Omran H, Leigh MW, Dell S, Morgan L, Molina PL, et al. Congenital heart disease and other heterotaxic defects in a large cohort of patients with primary ciliary dyskinesia. Circulation (2007)

- 115:2814–21. 10.1161/CIRCULATIONAHA.106.649038 [PubMed] [CrossRef] [Google Scholar]
- Knowles MR, Daniels LA, Davis SD, Zariwala MA, Leigh MW. Primary ciliary dyskinesia. Recent advances in diagnostics, genetics, and characterization of clinical disease. Am J Respir Crit Care Med (2013) 188:913–22. 10.1164/rccm.201301-0059CI [PMC free article] [PubMed] [CrossRef] [Google Scholar]
- Fliegauf M, Benzing T, Omran H. When cilia go bad: cilia defects and ciliopathies. Nat Rev Mol Cell Biol (2007) 8:880–93. 10.1038/nrm2278 [PubMed] [CrossRef] [Google Scholar]
- Kuehni CE, Frischer T, Strippoli MP, Maurer E, Bush A, Nielsen KG, et al. Factors influencing age at diagnosis of primary ciliary dyskinesia in European children. Eur Respir J (2010) 36:1248–58. 10.1183/09031936.00001010 [PubMed] [CrossRef] [Google Scholar]
- Lucas JS, Walker WT, Kuehni CE, Lazor R. Primary ciliary dyskinesia. In: Courdier J-F, editor. Orphan Lung Diseases (Vol. 54), Lausanne: ERS Monograph; (2011). P. 201–17. [Google Scholar]
- O'Callaghan C, Chetcuti P, Moya E. High prevalence of primary ciliary dyskinesia in a British Asian population. Arch Dis Child (2010) 95:51–2. 10.1136/adc.2009.158493 [PubMed][CrossRef] [Google Scholar]
- Coren ME, Meeks M, Morrison I, Buchdahl RM, Bush A. Primary ciliary dyskinesia: age at diagnosis and symptom history. Acta Paediatr (2002) 91:667–9. 10.1111/j.1651- 2227.2002.tb03299.x [PubMed] [CrossRef] [Google Scholar]
- Sommer JU, Schäfer K, Omran H, Olbrich H, Wallmeier J, Blum A, et al. ENT manifestations in patients with primary ciliary dyskinesia: prevalence and significance of otorhinolaryngologic co-morbidities. Eur Arch Otorhinolaryngol (2011) 268:383–8. 10.1007/s00405-010-1341-9 [PubMed] [CrossRef] [Google Scholar]
- Werner C, Lablans M, Ataian M, Raidt J, Wallmeier J, Große-Onnebrink J, et al. An international registry for primary ciliary dyskinesia. Eur Respir J (2016) 47:849–59. 10.1183/13993003.00776-2015 [PubMed] [CrossRef] [Google Scholar]
- Werner C, Onnebrink JG, Omran H. Diagnosis and management of primary ciliary dyskinesia. Cilia (2015) 4:2. 10.1186/s13630-014-0011-8 [PMC free article] [PubMed][CrossRef] [Google Scholar]
- Fedakar A, Aydogdu C. Clinical features of neonates treated in the intensive care unit for respiratory distress. Turk J Pediatr (2011) 53:173–9. [PubMed] [Google Scholar]
- Ferkol T, Leigh M. Primary ciliary dyskinesia and newborn respiratory distress. Semin Perinatol (2006) 30:335–40. 10.1053/j.semperi.2005.11.001 [PubMed] [CrossRef] [GoogleScholar]
- Mullowney T, Manson D, Kim R, Stephens D, Shah V, Dell S. Primary ciliary dyskinesia and neonatal respiratory distress. Pediatrics (2014) 134:1160–6. 10.1542/peds.2014-0808 [PMC free article] [PubMed] [CrossRef] [Google Scholar]
- Fretzayas A, Moustaki M. Clinical spectrum of primary ciliary dyskinesia in childhood. World J Clin Pediatr. (2016) 5:57–62. 10.5409/wjcp.v5.i1.57 [PMC free article] [PubMed][CrossRef] [Google Scholar]
- Bush A, Hogg C. Primary ciliary dyskinesia: recent advances in epidemiology, diagnosis, management and relationship with the expanding spectrum of ciliopathy. Expert Rev Respir Med (2012) 6:663–82. 10.1586/ers.12.60 [PubMed] [CrossRef] [Google Scholar]
- Noone PG, Leigh MW, Sannuti A, Minnix SL, Carson JL, Hazucha M, et al. Primary ciliary dyskinesia: diagnostic and phenotypic features. Am J Respir Crit Care Med (2004) 169:459–67. 10.1164/rccm.200303-365OC [PubMed] [CrossRef] [Google Scholar]
- Oktem S, Karadag B, Erdem E, Gokdemir Y, Karakoc F, Dagli E, et al. Sleep disordered breathing in patients with primary ciliary dyskinesia. Pediatr Pulmonol (2013) 48:897–903. 10.1002/ppul.22710 [PubMed] [CrossRef] [Google Scholar]

- Santamaria F, Esposito M, Montella S, Cantone E, Mollica C, De Stefano S, et al. Sleep disordered breathing and airway disease in primary ciliary dyskinesia. Respirology (2014) 19:570–5. 10.1111/resp.12273 [PubMed] [CrossRef] [Google Scholar]
- Pifferi M, Bush A, Caramella D, Di Cicco M, Zangani M, Chinellato I, et al. Agenesis of paranasal sinuses and nasal nitric oxide in primary ciliary dyskinesia. Eur Respir J (2011) 37:566–71. 10.1183/09031936.00068810 [PubMed] [CrossRef] [Google Scholar]
- Montella S, Corcione A, Santamaria F. Recurrent pneumonia in children: a reasoned diagnostic approach and a single centre experience. Int J Mol Sci (2017) 18(2):E296. 10.3390/ijms18020296 [PMC free article] [PubMed] [CrossRef] [Google Scholar]
- Kumar A, Lodha R, Kumar P, Kabra SK. Non-cystic fibrosis bronchiectasis in children: clinical profile, etiology and outcome. Indian Pediatr (2015) 52:35–7. 10.1007/s13312-015- 0563-8 [PubMed] [CrossRef] [Google Scholar]
- Kim HY, Kwon JW, Seo J, Song YH, Kim BJ, Yu J, et al. Bronchiectasis in children: 10-year experience at a single institution. Allergy Asthma Immunol Res (2011) 3:39–45. 10.4168/aair.2011.3.1.39 [PMC free article] [PubMed] [CrossRef] [Google Scholar]
- Eastham KM, Fall AJ, Mitchell L, Spencer DA. The need to redefine non-cystic fibrosis bronchiectasis in childhood. Thorax (2004) 59:324–7. 10.1136/thx.2003.011577 [PMC free article] [PubMed] [CrossRef] [Google Scholar]
- Brown DE, Pittman JE, Leigh MW, Fordham L, Davis SD. Early lung disease in young children with primary ciliary dyskinesia. Pediatr Pulmonol (2008) 43:514–6. 10.1002/ppul.20792 [PubMed] [CrossRef] [Google Scholar]
- Kennedy MP, Noone PG, Leigh MW, Zariwala MA, Minnix SL, Knowles MR, et al. High- resolution CT of patients with primary ciliary dyskinesia. AJR Am J Roentgenol (2007) 188:1232–8. 10.2214/AJR.06.0965 [PubMed] [CrossRef] [Google Scholar]
- Santamaria F, Montella S, Tiddens HA, Guidi G, Casotti V, Maglione M, et al. Structural and functional lung disease in primary ciliary dyskinesia. Chest (2008) 134:351–7. 10.1378/chest.07-2812 [PubMed] [CrossRef] [Google Scholar]
- Brenner DJ, Hall EJ. Computed tomography – an increasing source of radiation exposure. N Engl J Med (2007) 357:2277–84. 10.1056/NEJMra072149 [PubMed] [CrossRef] [Google Scholar]
- Montella S, Santamaria F, Salvatore M, Pignata C, Maglione M, Iacotucci P, et al. Assessment of chest high-field magnetic resonance imaging in children and young adults with noncystic fibrosis chronic lung disease: comparison to high-resolution computed tomography and correlation with pulmonary function. Invest Radiol (2009) 44:532–8. 10.1097/RLI.0b013e3181b4c1ba [PubMed] [CrossRef] [Google Scholar]