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Review Article

Physiology Section

Kartagener's Syndrome: A Narrative Review on its Clinical Implications and Management

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ABSTRACT

Kartagener's Syndrome, alternatively known as Primary Ciliary Dyskinesia (PCD) is a rare and intricate inherited disorder that impacts the structure and function of cilia, leading to compromised mucociliary clearance. The defining features of this condition consist of chronic sinusitis, situs inversus totalis, and bronchiectasis, forming a characteristic triad. Kartagener's Syndrome presents numerous challenges in its diagnosis and management, and its clinical implications have significant ramifications for affected individuals. This review aims to comprehensively review and analyse the clinical impact of this syndrome, focusing on the diagnosis, treatment, and management modalities. The ciliary dysfunction in Kartagener's Syndrome disrupts the mucus clearance and pathogens from the respiratory tract, resulting in chronic infections, progressive lung damage, and respiratory failure in severe cases. Through a narrative review of literature, reviews, and case studies, the authors have explored clinicians' diagnostic challenges and the advances in genetic testing methods that aid in early and accurate diagnosis. The authors have discussed the multidisciplinary approach to manage Kartagener's Syndrome, which involves respiratory therapies, antibiotics, and surgical interventions to improve the patient's quality of life and prevent complications. The review paper also includes in-depth case studies of individuals with Kartagener's Syndrome, highlighting the variability in clinical presentations and treatment outcomes. These case studies provide valuable insights into the diverse manifestations of this syndrome, further enhancing the existing understanding of the disease and its management. Ultimately, this review is directed to raise awareness among healthcare professionals about Kartagener's Syndrome, its clinical implications, and its diagnostic and treatment challenges. By shedding light on this rare and intricate genetic disorder, the authors here promote early recognition and proactive management, leading to improved outcomes and better quality of life for individuals with Kartagener's Syndrome.

Keywords: Bronchiectasis, Cilia, Primary ciliary dyskinesia, Respiratory tract infections, Situs inversus

INTRODUCTION

Kartagener's Syndrome is a highly uncommon autosomal recessive genetic disorder affecting cilia [1]. This congenital malformation is characterised by a triad of situs inversus, chronic sinusitis, and bronchiectasis [2]. Situs inversus is characterised by the organs in the chest and abdomen being positioned in a mirror-image arrangement, as opposed to the usual human anatomy. Chronic sinusitis results from prolonged inflammation and swelling of the sinuses, which are the spaces inside the nose and head. Bronchiectasis occurs due to the widening of the airways of the lungs, which in turn leads to the building-up of excess mucus, increasing the vulnerability of the lungs to infection. Kartagener's Syndrome belongs to the PCDs subgroup, collectively called ciliopathies. This condition leads to impaired ciliary action throughout the body [3]. They either do not move at all or do not move properly. It affects the sperm cells, epithelial cells in the bronchi and oviducts, and ependymal cells that line brain vesicles. In 1933, Kartagener recognised the causative relationship between the triad elements. This Syndrome is more common in consanguineous marriages [4].

Cilia are slender thread-like projections extending from the cell membrane. They are involved in diverse biological functions. Cilia can be both sensory and motile. Primary defects in their structure and process result in multiple ciliopathies. The most prominent is Kartagener's Syndrome. The primary functions of cilia are participating in the primary innate defense mechanism within the upper respiratory tract by facilitating mucociliary clearance, and safeguards the lungs from the detrimental effects of allergens, inhaled pollutants, and pathogens. Also, helps move mucous and other vital substances, and plays a critical role in establishing and maintaining the left-right axis during embryogenesis, In the lumen of the uterine tube, it generates currents that propel the egg cell toward the uterus, thus facilitating fertilisation in the appropriate area [5]. The dysfunction of cilia causes the accumulation of mucous and the colonisation of bacteria in the upper portion of the respiratory tract [4].

Symptoms Associated with Kartagener's Syndrome

Consequently, this condition gives rise to prominent symptoms. The severity of symptoms generally varies from person to person. Symptoms associated with abnormal cilia and their location has been shown in [Table/Fig-1] [6].

S. No.	Author name	Location	Symptoms associated with abnormal cilia
1)	Mathur P [6]	**Upper respiratory tract**	1. Frequent upper and lower respiratory tract infections 2. Rhinitis 3. Persistent cough
2)		**Lower respiratory tract**	1. Frequent lung infections 2. Chronic coughing 3. Excess mucus 4. Frequent sinus infections 5. Middle ear infections 6. Loss of hearing 7. Idiopathic pulmonary fibrosis 8. Drastic lung damage
3)		**Brain**	1. Hydrocephalus

[Table/Fig-1]: Symptoms associated with abnormal cilia and their location [6].

Common associations include complex heart disease, polysplenia, and biliary atresia [6]. In men, infertility is caused by asthenozoospermia (reduced or absence of mobility of sperm). It may also be due to dysfunctional cilia in the epididymal duct [7]. In women, ectopic pregnancies might occur since the egg fails to reach the site of fertilisation and gets implanted at locations other than the uterus [8].

The initial manifestation of this syndrome emerges during the neonatal period. Newborns affected by the condition may experience coughing, choking, gagging, and respiratory distress shortly after birth [9]. These events mainly occur due to improper movement of cilia since they are required to clear a newborn's lungs from amniotic fluid acquired from the womb, if present [9]. The diagnosis often goes unnoticed because the symptoms are non-specific, and there is limited awareness of the disease in primary care settings. Furthermore, more than the available tests are individually accurate enough for a definitive diagnosis. These tests require expensive and sophisticated equipment and expertise for proper analysis and interpretation, making diagnosis accessible only at highly specialised centers. The challenge is even more significant in countries with limited resources due to the scarcity of such costly equipment and technical knowledge [10]. This review aimed to shed light on the necessity for a multidisciplinary approach required for managing Kartagener's Syndrome.

Clinical Investigations

Currently, there is no universally standardised algorithm defining the criteria for diagnosis, and there are no international guidelines for conducting and reporting tests. Most specialised centers rely on "local" diagnostic algorithms [10]. Following a European consensus statement, the diagnosis of PCD should be established by observing a phenotype consistent with the disease and validating it through diagnostic tests conducted at specialised centers [4]. Although there is conflicting information within the existing literature, specific anatomical alterations identifiable through Computed Tomography (CT) scans of the paranasal sinuses have been proposed as potential factors influencing the onset of Chronic Rhinosinusitis (CRS). Notably, the existence of anatomical variations such as Concha Bullosa (CB), Paradoxical Middle Turbinate (PMT), infraorbital ethmoid (Haller) cells, fronto-ethmoidal cells, suprabullar cells, and supraorbital cells may impede proper drainage from the frontal sinus or the ostiomeatal complex [11]. Clinical investigations have been represented in [Table/Fig-2] [4,12].

S. No.	Diagnostic tests	Clinical manifestation of Kartagener syndrome	Highlighted features of the test
1)	• Transmission Electron Microscopy (TEM) (The best method to investigate the condition) • X-ray • CT scan • Hearing tests • Nitric oxide tests • High-Speed Video Microscopy (HSVM) • Ciliary culture Kugner M [12].	Ciliary defects	Transmission Electron Microscopy (TEM) is used to detect ciliary ultrastructural defects Kugner M [12].
2)	• Nasal nitric oxide (nno) • High-Speed Video Microscopy (HSVM), • Transmission Electron Microscopy (TEM), • Immunofluorescence (IF) • Genetic testing Olm MAK et al.	Ciliary defects	High-speed video imaging enables the visualisation of the typical ciliary beating pattern. Ciliary beat frequency and pattern aid in comprehending the impact of ciliary abnormalities on mucus movement. Olm MAK et al., [4].

[Table/Fig-2]: Clinical investigations [4,12].

Differential Diagnosis

Cystic Fibrosis (CF), immunodeficiency syndromes, and gastroesophageal reflux are the primary differential diagnoses for this condition [13]. Others are Adenoid hyperplasia, Alpha1-Antitrypsin (AAT) Deficiency, Bronchial obstruction, Chronic aspiration (COPD), Congenital cartilage deficiency, CF, foreign body aspiration, idiopathic interstitial pneumonia, idiopathic nasal polyposis, inhalation of toxic substances, interstitial lung diseases, including idiopathic pulmonary fibrosis, tracheobronchomegaly [14].

Diagnosis

Abnormal ciliary function may influence the positioning of internal organs, with genetic factors shaping embryonic protein structures that guide organ development. Proteins like Sonic Hedgehog and Nodal play roles in left-sided development, while others like Activin beta are involved on the right. Disruptions in these processes can lead to misplacements [15]. Conditions like PCD often result in situs inversus totalis (complete organ reversal) or situs ambiguous (irregular organ laterality) [16]. This complexity is furthered by differing interpretations among specialists, particularly cardiologists, who associate heterotaxy with specific heart anomalies. Heterotaxy occurs in about one in 10,000 cases and involves various inheritance patterns [17,18]. Dysfunctional cilia in embryonic cells contribute to these laterality abnormalities and heart defects [16].

Diagnosing this syndrome can be challenging due to its non-specific symptoms, such as recurrent respiratory infections and chronic cough, leading to delays in diagnosis [12]. There's no gold standard test, so diagnosis relies on a thorough medical history, physical examination, and a battery of tests [12]. In specific communities, the social stigma surrounding the diagnosis of a chronic illness or genetic disease acts as a barrier that deters individuals from seeking medical assistance. The association of such conditions with infertility further contributes to the stigma's impact [10]. Social stigma, particularly around chronic and genetic diseases, can hinder individuals from seeking medical help, especially when coupled with infertility concerns. Patients typically present with symptoms like cold, coughing, shortness of breath, expectoration, anosmia, headaches, and childhood complaints like frequent colds, ear discharge, and nasal congestion [19,20]. This disorder may be under-diagnosed in minority populations. In chronic respiratory diseases like PCD and Secondary Ciliary Dyskinesia (SCD), elevated levels of inflammatory proteins like Matrix Metalloproteinases (MMPs) contribute to airway inflammation and remodeling, starting early and progressing unpredictably [21].

Treatment and Management

While there is no cure for this syndrome, the primary focus of treatment revolves around maintaining clear airways and facilitating the removal of thick, excess fluid, mucous, and debris. This therapeutic approach is called airway clearance therapy [22].

Infertility in males with Kartagener Syndrome results from the absence of sperm motility. At the same time, in females, it arises due to impaired ovum transport attributed to the dyskinetic motion of the oviductal cilia. Overall, the prognosis for these patients tends to be poor, even with treatment. In some cases, the patient may have an elevated risk of tuberculosis. Tuberculosis frequently complicates lower lobe bronchiectasis. Sometimes, surgical intervention would not yield improvement if bronchiectasis is present in both lungs. The management of recurrent infections should focus on antibiotic control. While some reports suggest sinus surgery as a viable option for such patients with tuberculosis, this remains an area necessitating further investigation [23].

Non-surgical

The medical management for an individual with Kartagener Syndrome centers around preventing respiratory infections [19]. Several methods, encompassing manual chest physiotherapy, postural drainage, autogenic drainage, active cycle breathing, and exercise, are employed to ensure efficient airway clearance [24]. Non-surgical treatment measures mainly include the use of antibiotics, steroids, nebulisation, and nasal decongestants. Other conservative management techniques include regular chest physiotherapy, antibiotics that provide adequate pseudomonal coverage, and

supportive pulmonary care. The usefulness of DNase and other mucolytic medicines such as hypertonic saline and acetylcysteine has not been adequately examined in these individuals but may be explored, especially in patients with recurrent infections or chronic respiratory problems [25].

Surgical treatment

Understanding the precise arrangement of abdominal organs and the branching pattern of primary stem bronchi is crucial for classifying malpositions and guiding surgical interventions. Dextrocardia with situs inversus typically presents the heart in the expected position, whereas dextrocardia without situs inversus, cases of situs ambiguus, or isolated levocardia often involve complex, multiple anomalies. Dextrocardia with complete situs inversus occurs in approximately two per 10,000 births, with congenital heart disease being relatively rare at around 3% of cases, while Kartagener's Syndrome may affect about 20% of these patients. The overall incidence is estimated to be roughly one in 50,000 births. Cases associated with surgical intervention are given below in [Table/Fig-3] [26-30].

The cases discussed are lung transplantation [26], Laparoscopic sleeve gastrectomy with Kartagener Syndrome [27]. A case of Kartagener Syndrome with Rhinolalia Clausa [28]. Endoscopic sinus surgery for treatment of Kartagener syndrome [29], extracorporeal life support in Kartagener's Syndrome [30].

Cases associated with Kartagener's Syndrome in the recent past have been tabulated under [Table/Fig-4] [31-33].

Complications

Complications in patients with Kartagener's Syndrome are given below in [Table/Fig-5] [34-37].

Ref. No.	Author name and year	Age of patient	Complications	Type of surgical management	Outcome
[26]	Yamamoto H, et al., 2021	46 years	Adrenocorticotropic hormone deficiency	Lung transplantation with lobar reduction	The atelectasis of the right lower lobe resolved, and the transplanted lung expanded to fill the space in the cardiac notch of the left lung.
[27]	Burvill A, et al., 2019	23 years	Morbid obesity	Laparoscopic sleeve gastrectomy	The patient experienced a notable weight reduction and demonstrated good tolerance to nourishing fluids.
[28]	Raoufi M et al., 2016	18 years	Nasal polyposis and co-existing sinusitis	Management was non-surgical. But similar cases may require Endoscopic sinus surgery including the creation of a nasal antral window beneath the inferior turbinate	After 6 months of treatment swelling subsided and no signs of local recurrence or other tuberculous location.
[29]	Tang X et al., 2013	17 years	Bronchiectasis and situs inversus	Underwent Functional Endoscopic Sinus Surgery (FESS)	The frequency of lung infection episodes decreased to one or two times a year, eliminating the need for hospitalisation. The severity and duration of airway infections were notably reduced.
[30]	Kundu R et al., 2022	43 years	Severe respiratory acidosis with hypoxemia, leading to unstable haemodynamic conditions	Venovenous- Extracorporeal Membrane Oxygenation (VV-ECMO) therapy Percutaneous tracheostomy	A subsequent High Resolution Computed Tomography (HRCT) chest revealed bilateral bronchiectatic changes accompanied by multifocal patchy areas of confluent alveolar densities/centrilobular nodules in both lung fields.

[Table/Fig-3]: Cases associated with surgical interventions [26-30].

Ref. No.	Author name and year	Age of the patient	Highlighted features	Type and details of management chosen	Outcome
[31]	Kara B et al., 2021	10 years	Bilateral multiple cervical lymphadenopathies Undifferentiated nasopharyngeal carcinoma.	Following chemotherapy comprising cisplatin, docetaxel, and 5-fluorouracil, significant regression of nasopharyngeal carcinoma and lymphadenopathies occurred after four cycles. Subsequent radiotherapy to the nasopharynx and cervical regions led to six years of tumour-free follow-up. Nonetheless, ongoing supervision by paediatric immunology and allergy departments persists due to recurrent respiratory infections and sinusitis.	Tumour-free status The patient continues to experience recurrent respiratory infections and sinusitis.
[32]	Sehgal IS, et al., 2015	31 years	Recurrent exacerbations characterised by increased cough, purulent expectoration, haemoptysis, and breathlessness. Additionally, nasal stuffiness, obstruction, and epistaxis have been reported since childhood.	Oral prednisolone at 0.5 mg/kg body weight, tapered over four months, and discontinued. Additionally, a combination of inhaled fluticasone (500 µg/day) and formoterol (12 µg/day) via pressurised metered-dose inhaler was administered.	Significant clinical and radiological improvement. decrease in serum IgE to 948 IU/mL. Repeat spirometry revealed no improvement in forced expiratory volume in 1 second or forced vital capacity values.
[33]	Sahu S et al., 2022	22 years	No prior history of allergies or weight loss was reported, nor was there any family history of atopy or asthma. Both parents were healthy, and the pregnancy was uneventful, occurring in a non-consanguineous marriage.	Supplemental oxygen with a simple face mask at 10 L/min Non-Invasive Ventilation (NIV) and intravenous antibiotics, systemic corticosteroids, inhaled bronchodilators, mucolytics, chest physiotherapy, and other supportive treatments.	Discharged on the seventh day, with recommendations for domiciliary oxygen therapy, oral antibiotics, and chest physiotherapy. Additionally advised on the importance of annual influenza and pneumococcal vaccinations, along with other essential preventive measures.

[Table/Fig-4]: Cases associated with Kartagener's syndrome in the recent past [31-33].

Complications in patients with Kartagener's Syndrome				
Author, year and reference number	**Age of patient and sex**	**Complications**	**Clinical presentation of the complication**	**Clinical investigation used for definitive diagnosis**
Oka K et al., 2018 [34]	36 years female	Ig A nephropathy (most prevalent form of primary chronic glomerulonephritis among other renal conditions) Patients with Kartagener's syndrome have shown a range of renal abnormalities, encompassing different glomerular injuries and polycystic kidney disease	Asymptomatic microhaematuria and proteinuria	Biopsy
Nagai Y et al., 2020 [35]	39 years, female (non-smoker)	Pulmonary mucoepidermoid carcinoma (relatively uncommon)	Lung cancer originated from the bronchus of the right upper lobe. Low-grade malignancy	Histological analysis

Masaki H et al., 1993 [36]	38 years male	Lung abscess	Recurrent pneumonia	Broncho-fiberscopy
Horie M et al., 2010 [37]	71 years male	Squamous cell carcinoma (Lung cancer)	Chronic sinusitis and deafness since childhood	Transbronchial lung biopsy of the right lung tumour

[Table/Fig-5]: Complications in patients with Kartagener's Syndrome [34-37].

COVID-19 PCD Correlation

The Coronavirus Disease-2019 (COVID-19) pandemic, driven by the Severe Acute Respiratory Syndrome Coronavirus 2 (SARS-CoV-2), had infected over 115 million individuals globally and resulted in more than 2.5 million deaths by March 2021. Those with pre-existing chronic health conditions are known to be particularly susceptible to contracting the disease and experiencing a severe disease progression, PCD. In March 2020, the University of Bern initiated a study to provide insights into the risk and progression of COVID-19 among individuals with PCD. This initiative resulted in the establishment of COVID-PCD, a longitudinal online survey focused on assessing the health status, protective measures, and quality of life of those with PCD throughout the pandemic [38]. The COVID-2019 primarily affects the respiratory system, giving rise to symptoms such as cough, fever, and fatigue. However, the virus can manifest with other symptoms affecting different body parts. Among these, an unexpected sign that has gained recognition among healthcare professionals and the general public worldwide is the loss of smell. Recent clinical research has revealed a strong connection between COVID-19 patients in the United States and the impairment of odor and taste. This association between the onset of chemosensory dysfunction (specifically, loss of smell and taste) and COVID-19 positivity has also been observed in Iran and Italy since the start of the pandemic. Hence, the loss of smell or taste might serve as subclinical indicators or potential early disease symptoms [39].

CONCLUSION(S)

Kartagener's Syndrome manifests as a rare autosomal recessive disorder typified by a triad of symptoms, namely situs inversus, chronic sinusitis, and bronchiectasis. It arises from abnormalities in ciliary structure or function, impairing mucociliary clearance and predisposing individuals to respiratory infections. Timely diagnosis through clinical evaluation, imaging, and genetic testing is crucial for effective management. Treatment focuses on symptom control, including antibiotics, airway clearance techniques, and surgery when necessary. Despite its rarity, Kartagener's Syndrome offers valuable insights into ciliary function and its implications for respiratory health. Further research is needed to advance therapeutic interventions and improve patient outcomes. A multidisciplinary approach is essential for comprehensive care. Increased awareness and early diagnosis are critical for enhancing the quality of life for affected individuals.

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