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Review Article
Kartagener's syndrome

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Kartagener's syndrome with Dextrocardia - A case report

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One rare variant of immotile cilia syndrome is known as Kartagener's syndrome. The sinus, eustachian tube, middle ear, and respiratory tract cilia are all impacted by Kartagener's syndrome. These cilia's inability or improper movement causes persistent issues with the ear and sinuses. Some individuals with dextrocardia may not experience any symptoms, while others may have heart-related issues or other associated health problems. Diagnosis typically involves imaging studies such as X-rays, echocardiograms, or CT scans to visualize the position of the heart and other organs. Treatment depends on the presence of any associated heart defects or health issues. In many cases, individuals with dextrocardia can lead normal, healthy lives with proper medical management. Close collaboration between patients and healthcare providers is essential for individuals with dextrocardia to track their heart health and general wellness. If needed, regular check-ups and appropriate medical interventions can help manage any potential complications associated with this condition. Individuals with dextrocardia need to work closely with healthcare professionals to monitor their heart health and overall well-being. If needed, regular check-ups and appropriate medical interventions can help manage any potential complications associated with this condition.

Keywords: Dextrocardia, Echocardiograms, Eustachian tubes

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Introduction

Immotile cilia syndrome, also known as Kartagener's Syndrome or PCD, is a hereditary condition that has no known cause that affects the cilia, which are microscopic hair-like structures that line the respiratory system, sinuses, eustachian tubes, middle ear, fallopian tubes, and both the upper and lower respiratory tracts [1].

The inability or improper movement of these cilia causes persistent issues with the ear, sinuses, and lungs in Kartagener's syndrome. Recurrent chest infections and symptoms affecting the nose, ears, and throat might be signs of decreased ciliary motility, which in turn causes infertility in this illness. Although estimates vary between 1 in 12,500 and 1 in 50,000, the approximate frequency of PCD is almost 1 in 30,000. two [2] Some key symptoms of Kartagener's syndrome include middle ear infections, hearing loss, infertility, persistent coughing, recurrent sinus infections, and frequent lung infections. A catastrophic prognosis may result in complex circumstances where bilateral lung transplantation is postponed [3].

Biopsies that confirm the reduced cilia function and genetic investigations can validate Kartagener's syndrome, the primary diagnosis. The major goal of supportive therapy is to keep the dextrocardia, genetic testing, and ciliary beat frequency clinical steps for diagnosis. Computerized tomography (CT) scans, X-rays, and audiological exams are some imaging procedures doctors may prescribe to detect problems [4]. The cilia that line the respiratory tract and fallopian tube are responsible for ciliary motility, which is reduced in Kartagener's disease. Because the cilia that line the airways aren't working correctly, mucociliary clearance is low. [5]

The right side of the thorax is where you'll find the heart, pointing to the right, in the sporadic congenital disease known as dextrocardia. [6] It is believed to be a result of the organs being in an aberrant position when the embryo is developing. However, symptoms like blue skin, trouble breathing, stunted growth and weight gain, lethargy, jaundice, pale skin, and recurrent sinus or lung infections might be caused by disorders related to dextrocardia. two [7] While most cases of isolated dextrocardia are harmless, symptoms such as cyanosis, dyspnea, and failure to grow can occur in babies.

Newborns and neonates may experience respiratory difficulties, recurrent infections, and failure to grow if it is present alongside other congenital defects. [8] and [7] Infertility may also be linked to dextrocardia in adults. Electrocardiograms, CT scans of the heart, and chest X-rays are some of the diagnostic tools doctors use to identify the illness. [7] Other congenital abnormalities impact treatment decisions; in extreme situations involving linked heart defects or problematic genetic disorders, surgery may be required. People who have uncomplicated dextrocardia should expect a normal life expectancy notwithstanding the possibility of complications [8].

Although there is currently no method to prevent dextrocardia, it is suggested that pregnant women refrain from using illicit substances, particularly cocaine, before and throughout their pregnancy. [8]

There are two main types of dextrocardia:

1. Dextrocardia with Situs Inversus Totalis (SIT): This is the most common form of dextrocardia. In addition to the heart being on the right side, the other organs in the chest and abdomen are also mirrored or reversed from their normal positions. Left to right, for instance, you'll find the liver and the spleen. "Situs inversus totalis" describes this extreme case of organ implantation inversion.

2. Dextrocardia without Situs Inversus (Isolated Dextrocardia): In this type, On the right side, you can only see the heart., while the other organs maintain their usual placement. This form is less common than dextrocardia with situs inversus. Although the precise reason behind dextrocardia is not always clear, it is believed to be caused by problems that occur throughout the process of fetal development. It may be associated with other congenital heart defects or genetic conditions. Some individuals with dextrocardia may not experience any symptoms, while others may have heart-related issues or other associated health problems.

Diagnosis typically involves imaging studies such as X-rays, echocardiograms, or CT scans to visualize the position of the heart and other organs. Treatment depends on the presence of any associated heart defects or health issues. In many cases, individuals with dextrocardia can lead normal, healthy lives with proper medical management.

Close collaboration between patients and healthcare providers is essential for individuals with dextrocardia to track their heart health and general wellness. If needed, regular check-ups and appropriate medical interventions can help manage any potential complications associated with this condition.

Individuals with dextrocardia need to work closely with healthcare professionals to monitor their heart health and overall well-being. If needed, regular check-ups and appropriate medical interventions can help manage any potential complications associated with this condition.

Case Report

The pediatric ward admitted a male boy, 13 years old, who complained of weakness, coughing, and loud breathing. When the patient first complained of his on/off cough, which he said was consistent with seasonal change, non-productive, and accompanied by loud breathing, he was perfectly healthy six years ago. A temperature of 98.5 degrees Fahrenheit, a heart rate of 92 beats per minute, and a respiratory rate of 22 breaths per minute were considered normal vital signs upon presentation.

The patient's hemoglobin level was 12.3 gm/dl when they were admitted.TLC was 18.9 thousand cu/mm. Aspartate transaminase range was 54 U/l and Alanine transaminase was 60U/L. Other investigations showed amylase was slightly high and Chloride was low. In transthoracic echocardiography, situs inversus, dextrocardia, and mild pericardial effusion were seen.

CECT manifested that situs inversus totalis with active infective etiology, cylindrical and cystic bronchiectasis involving the middle and lower lobes of a lung on the left side, and Moderate bilateral maxillary and ethmoid sinusitis and a CT scan of PNS was evident that bilateral maxillary and ethmoidal sinusitis and bilateral inferior turbinate hypertrophy.

The patient's skiagram chest PA view shows that the situs is inversus with dextrocardia. Ultrasonography of the abdomen revealed that situs inversus. During audiogram testing, it showed that mild hearing loss was seen at high frequency.

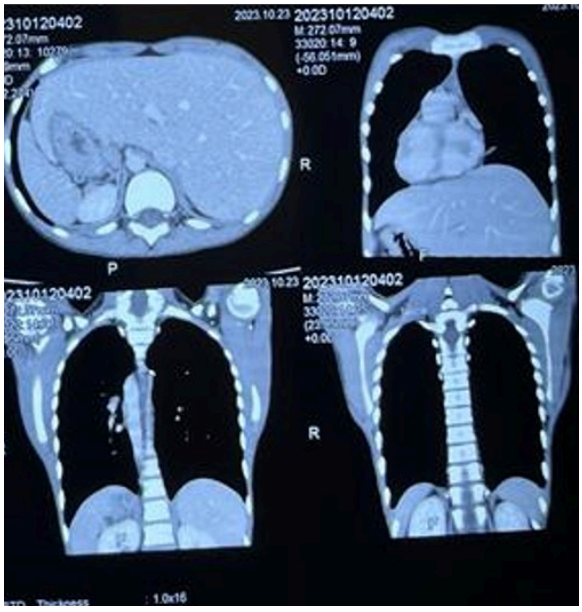

Fig 1: shows the (Situs inversus totalis with active infective etiology, cylindrical, and cystic bronchiectasis involving the middle and lower lobes of the lung on the left side. Moderate bilateral maxillary and ethmoid sinusitis- the possibility of Kartagener syndrome to be considered).

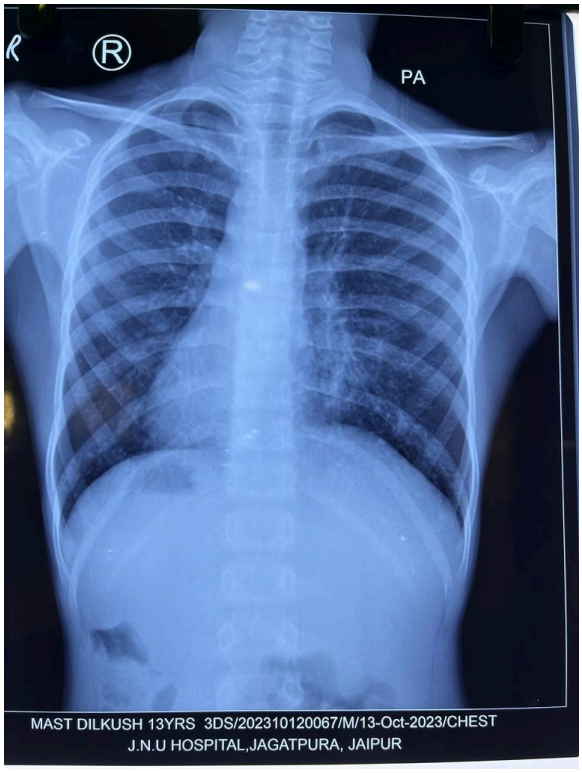

Fig 2: shows the (Cystic bronchiectasis changes in the lower and mid zones with dextrocardia).

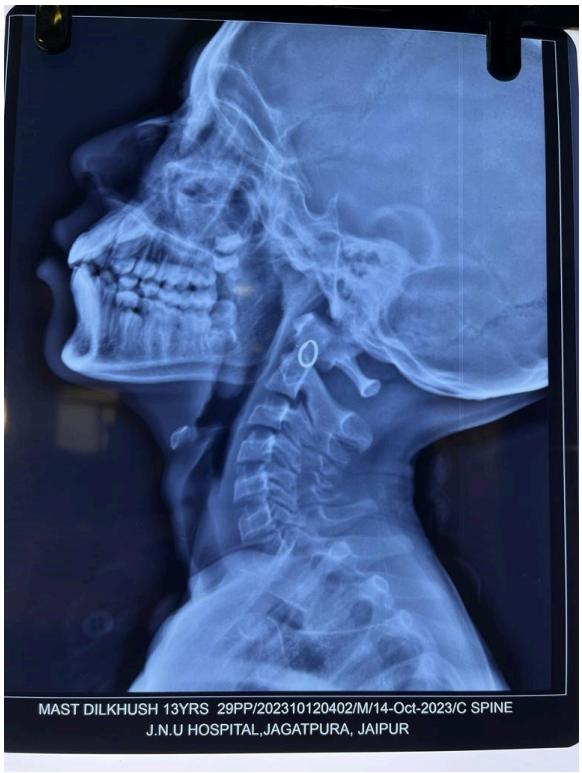

Fig 3: shows the (Bilateral maxillary and Ethmoidal Sinusitis & Bilateral Inferior Turbinate Hypertrophy).

Discussion

Prevalence estimates for Kartagener's syndrome PCD range from 1 in 12,500 to 1 in 50,000, with an estimated 1 in 30,000 cases. An ultrastructural genetic defect in KS hinders ciliary motility, which in turn causes recurrent infections of the sinuses, ear, nose, and throat (ENT), and the reproductive system. These young patients need access to treatment options for infertility as soon as feasible, thus it's important to diagnose them early whenever possible with a high index of suspicion. [9] The patient or sibling must have situs inversus, the spermatozoa must be alive but not motile, the transbronchial mucociliary clearance must be decreased or nonexistent, and electron microscopy must reveal cilia with a distinctive ultrastructural abnormality for this syndrome to be diagnosed. Recurrent chest infections, bronchitis, and rhinitis that have persisted from childhood should also be indicated by the clinical picture. [10] Dextrocardia is a rare congenital anomaly that may be associated with significant extracardiac problems; it affects an estimated 1 out of every 12,000 live births. There is an equal number of males to females. [11] On the other hand, dextrocardia with situs inversus is linked to a reduced incidence of congenital heart disease, 0–10%. Dextrocardia with a normal abdominal situs has a high incidence of related congenital cardiac defects. About 1 in 10,000 people have situs inversus totalis, which may be linked to primary ciliary dyskinesia (PCD), a condition in which cilia malfunction. [13] Studying ciliary motility (frequency and beat pattern) using high-resolution digital video and high speed, in conjunction with ciliary ultrastructure by electron microscopy, provides the basis for the diagnosis of primary ciliary dyskinesia (PCD). **[12]**

Conclusion

In this case report we discussed Kartagener's syndrome with Dextrocardia. Our study reports that her symptoms were relieved after taking the proper medication prescribed by the physician. can be a helpful diagnostic tool in detecting the condition. Early treatment is indicated for any further complications.

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