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Case Report

# Primary ciliary dyskinesia – Kartagner syndrome

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**Abstract**: PCD is an inherited disorder characterised by impaired ciliary function leading to chronic Sino pulmonary disease, persistent middle ear infections, left –right orientation defects and infertility. Estimated frequency of PCD is 1 in 12,000 to 1 in 20,000 live births. It is diagnosed by recurrent respiratory tract infections and presence of clinical phenotype and ultra-structural defects of cilia. We are reporting a case of kartagner syndrome in a 12 yrs. old child. She had history of recurrent respiratory tract infection, chronic otitis media. Kartagner was diagnosed by phenotypic presentation and HRCT THORAX suggestive of dextrocardia with situs in versus with bronchiectasis with recurrent Sino pulmonary disease and persistent middle ear infection.

**Key words**: kartagner syndrome; situs in versus; chronic sinusitis and bronchiectasis

## Introduction

Kartagner syndrome is an inherited disorder characterized by chronic Sino pulmonary disease, persistent middle ear effusions, laterality defects and infertility. Estimated frequency of PCD is 1 in 12,000 to 1 in 20,000, its prevalence in children with repeated respiratory infections has been estimated to be as high as 5%. Diagnosed by presence of clinical phenotype, imaging studies, pulmonary function tests and ultra-structural defects of cilia.

**Case study:**

We are reporting a case of Kartganer syndrome which has incidence of 1 in 12,000 to 1 in 20,000. A 12 years old female child brought to our tertiary care center with complaints of recurrent respiratory tract infection since birth, ear discharge since past 5 years, and difficulty in breathing since past 1 month. History of frequent hospital visits for recurrent respiratory illness for which child was treated symptomatically. On admission child was having temperature of 101 F in right axilla, pulse rate 110 b/min, resp rate 26 cycles/min, BP-116/70 mmHg, spo2 88-90%.

**ENT examination**:

Bilateral ear discharge present with large central perforation in left ear, features suggestive of chronic otitis media.

**On systemic examination**:

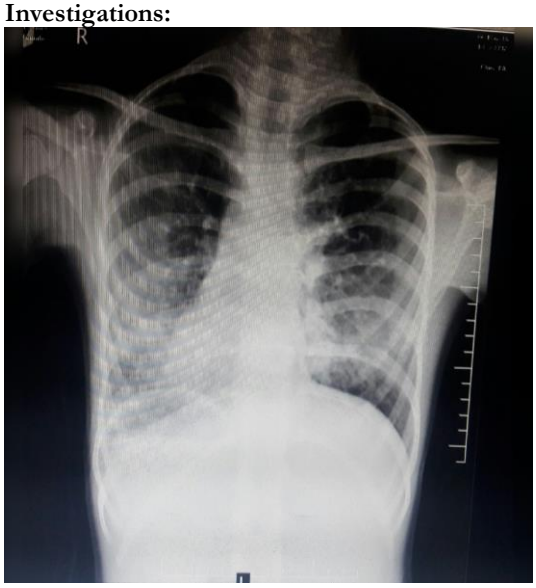
**Respiratory system**

Air entry decreased on both sides with bilateral crept, increased work of breathing, subcostal retractions present

**Cardiovascular system**:

Apex beat noted towards right 4 intercostal space, S1S2 heard. Per abdomen and CNS examination did not reveal any abnormality.

**Investigations:**



**Chest Xray**

Stomach gas shadow on right side with inhomogenous radiopacities in bilateral lower lung zones.

**USG Abdomen and Pelvis**

Spleen and stomach appears on right side; liver and gall bladder appear on left side. Features suggestive of situs in versus.

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**2D ECHO**
Dextrocardia with good biventricular function.

**HRCT Thorax**
Dextrocardia with situs in versus

1. Areas of cystic and cylindrical bronchiectasis in upper and middle lobe of left lung and lingular lobe of right lung
2. Multiple tiny peribroncho –vascular nodules in basal segments of right lower lobe indicate active pulmonary infection.

**Ear Swab Culture and Sensitivity**
Acinetobacter species isolated.

## Conclusion

Kartagners syndrome is an inherited disorder that predisposes to repeated respiratory tract infections with clinical manifestations of lower respiratory tract disease tend to increase with age and become the leading cause of morbidity and mortality. Regular spirometry; chest imaging and sputum or oropharyngeal cultures to be done to slow down the progression and extent of lung diagnosis by early diagnosis and treatment.

## References

1. Thomas W. Ferkal Jr. "Primary Ciliary Dyskinesia (Kartagners Syndrome) Nelson Textbook of Pedatrics 2.20(15) 2113-2116. Elsevier Publications.

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