

Delayed Diagnosis of Kartagener Syndrome in an Adolescent with Recurrent Sinopulmonary Infections and Severe Bronchiectasis: The First Reported Case from Somalia

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Case Report

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Abstract

Background

Kartagener syndrome is a rare form of primary ciliary dyskinesia characterized by chronic sinopulmonary disease, bronchiectasis, and situs inversus. Diagnosis is frequently delayed because respiratory manifestations are nonspecific and specialized diagnostic testing may be unavailable, particularly in resource-limited settings. We report what is, to our knowledge, the first reported case of Kartagener syndrome from Somalia, highlighting the diagnostic and management challenges encountered in a resource-limited setting.

Case presentation

A 15-year-old girl from rural Somalia presented with a lifelong history of recurrent sinopulmonary infections, chronic productive cough, progressive exertional dyspnea, and intermittent hemoptysis. Examination revealed digital clubbing and signs of chronic respiratory disease. Chest radiography and contrast-enhanced computed tomography demonstrated dextrocardia, situs inversus totalis, and extensive bilateral cystic bronchiectasis with superimposed bronchopneumonia. A clinical diagnosis of Kartagener syndrome was made based on the characteristic phenotype, radiological findings, and a PICADAR score of 8/14. Confirmatory investigations, including ciliary function studies and genetic testing, were unavailable because of resource limitations. She was treated with intravenous antibiotics, bronchodilator therapy, and structured airway-clearance therapy, followed by oral azithromycin and daily home-based chest physiotherapy. Family education focused on airway-clearance techniques, recognition of respiratory exacerbations, treatment adherence, and long-term follow-up. Because of the substantial geographic distance from specialist care, follow-up was coordinated with a local primary care physician and supported by telemedicine. At 1 month, her cough and sputum production had markedly decreased, respiratory distress had resolved, and she had returned to school and daily activities.

Conclusions

Kartagener syndrome should be considered in children and adolescents with recurrent sinopulmonary infections, chronic rhinosinusitis, bronchiectasis, and laterality abnormalities. In resource-limited settings, characteristic clinical and radiological findings can facilitate recognition when definitive diagnostic testing is unavailable, although the limitations of a clinical diagnosis should be acknowledged. Early recognition, sustained airway-clearance therapy, appropriate treatment of respiratory infections, and patient- and family-centered education are important for long-term disease management. This case also illustrates how locally coordinated and remote follow-up may help maintain continuity of care when access to specialized respiratory services is limited.

Introduction

Kartagener syndrome (KS) is a rare autosomal recessive disorder and a subtype of primary ciliary dyskinesia (PCD), a group of inherited disorders caused by impaired motile ciliary function [1]. It is classically characterized by the triad of chronic sinusitis, bronchiectasis, and situs inversus [2]. The association between these clinical features was first described by Siewert in 1904, and the syndrome was later recognized as a distinct congenital disorder by Manes Kartagener in 1933 [2, 3]. The reported incidence ranges from approximately 1 in 30,000 to 1 in 40,000 live births [1, 3].

KS results from congenital defects affecting the structure or function of motile cilia, leading to impaired mucociliary clearance in the respiratory tract and reduced sperm motility in males. During embryonic development, normal ciliary function is also essential for establishing left-right body asymmetry [1, 4]. Mutations in the *DNAH5* and *DNAI1* genes are among the recognized genetic causes and are associated with defects of the outer dynein arms, resulting in impaired ciliary movement [4, 6]. As a result, patients are predisposed to recurrent upper and lower respiratory tract infections, while disruption of left-right axis formation leads to situs inversus in approximately 50% of individuals with PCD. The clinical presentation is highly variable and often overlaps with other chronic respiratory diseases, contributing to delayed diagnosis [5–7].

In resource-limited settings, the diagnosis of PCD may be particularly challenging because recurrent respiratory infections are common, access to specialized diagnostic testing is limited, and chronic respiratory symptoms may be attributed to more prevalent infectious diseases. Consequently, patients with a characteristic PCD phenotype may remain unrecognized for years, allowing progressive and potentially irreversible bronchiectatic lung damage to develop. This diagnostic challenge is particularly relevant when laterality abnormalities are not recognized early or when previous episodes have been treated as recurrent respiratory infection or tuberculosis.

We report the case of a 15-year-old girl from Baidoa, Somalia, with a lifelong history of recurrent sinopulmonary infections who remained undiagnosed until adolescence and presented with advanced bilateral cystic bronchiectasis and complete situs inversus totalis. To our knowledge, this is the first reported case of Kartagener syndrome from Somalia. The case highlights how integration of clinical history, recognition of laterality abnormalities, and characteristic radiological findings can facilitate diagnosis when definitive PCD testing is unavailable, and how family-centered airway-clearance training and decentralized follow-up may provide a practical approach to long-term management in resource-limited settings.

Case Presentation

A 15-year-old girl from Baidoa, Bay Region, Somalia, was brought to the emergency department by her mother with worsening productive cough, progressive shortness of breath, and intermittent hemoptysis.

She was born at term by spontaneous vaginal delivery without neonatal complications. Her developmental milestones were achieved appropriately, although recurrent illnesses subsequently affected her school attendance. According to her mother, recurrent respiratory symptoms began in early

childhood, characterized by repeated episodes of productive cough and lower respiratory tract infections requiring multiple hospital admissions and repeated courses of oral and intravenous antibiotics. Despite treatment, the episodes recurred, and the cough gradually became persistent. Over time, she developed daily production of thick yellowish-green sputum.

During childhood, she also developed recurrent sinonasal symptoms, including chronic nasal obstruction and episodes of sinusitis. In 2019, she underwent nasal polypectomy for symptomatic nasal polyps, but nasal symptoms subsequently recurred. She also reported several years of anosmia and recurrent headaches associated with episodes of sinus congestion. In addition, she had a longstanding history of recurrent otitis media, associated with intermittent ear discharge and temporary hearing impairment during acute episodes. Her mother had noted a gradual decline in her hearing over time.

As she progressed into adolescence, her respiratory symptoms became increasingly severe. She developed progressive exertional dyspnea, initially occurring only with vigorous physical activity but eventually limiting her ability to walk short distances and participate in sports with her classmates. This was accompanied by poor exercise tolerance and progressive fatigue. During the year preceding the current presentation, she experienced unintentional weight loss despite a preserved appetite, and her mother noticed progressive enlargement of her fingertips consistent with digital clubbing.

She also reported intermittent hemoptysis during the months preceding admission, usually presenting as blood-streaked sputum. On one occasion, she expectorated approximately one tablespoon of bright red blood. There was no history of massive hemoptysis.

During the six months preceding admission, she additionally experienced intermittent diffuse abdominal pain associated with abdominal distension and chronic constipation. The pain was unrelated to meals and was occasionally relieved following defecation. She also reported episodes of coughing while swallowing food or liquids but denied choking episodes.

She attained menarche at the expected age and reported regular menstrual cycles associated with dysmenorrhea. She had received routine childhood immunizations according to the national immunization schedule.

Her past medical history [Table 3] included treatment for pulmonary tuberculosis several years earlier and multiple courses of antibiotics for recurrent respiratory infections. She had previously undergone nasal polypectomy in 2019. She had no known drug allergies.

She was born to consanguineous parents who were first cousins. There was no known family history of similar chronic respiratory disease, primary immunodeficiency, or other inherited disorders. She lived with her parents and siblings and had never smoked or been exposed to household tobacco smoke. The family resided in a rural area of Somalia with limited access to specialized healthcare services. There was no recent history of travel.

During the two weeks before the current admission, her respiratory symptoms worsened considerably. Her productive cough became more frequent and severe, with increasing amounts of yellow-green sputum and intermittent bright-red blood staining. The worsening cough disturbed her sleep and further limited her daily activities. Her exertional dyspnea also progressed, prompting her mother to bring her to the emergency department for evaluation.

On admission, the patient appeared chronically ill, pale, and mildly cachectic. She was conscious, alert, and fully oriented to time, place, and person. Her weight was 27 kg, and her height was 130 cm. She was in mild-to-moderate respiratory distress with increased work of breathing and preferred to remain in a propped-up sitting position because of orthopnea. Digital clubbing was present in both hands. There was no cyanosis, jaundice, or peripheral edema.

Head and neck examination revealed tenderness over both maxillary sinuses. Nasal examination demonstrated congested nasal mucosa with a small recurrent right-sided nasal polyp. No cervical lymphadenopathy was detected. Respiratory examination demonstrated mildly reduced bilateral chest expansion with a centrally positioned trachea. Auscultation revealed widespread coarse bilateral crackles, predominantly over the lower lung fields, scattered rhonchi, and reduced air entry bilaterally. Cardiovascular examination demonstrated the apex beat in the right fifth intercostal space along the midclavicular line. Heart sounds were normal without audible murmurs. Abdominal examination revealed mild distension with generalized tenderness. The liver was palpable below the left costal margin. No splenomegaly or ascites was appreciated clinically, and bowel sounds were normal. Neurological examination was unremarkable, with no focal neurological deficits.

Chest radiography (CXR) demonstrated dextrocardia with bilateral bronchiectatic changes along both the right and left cardiac borders [Figure 1]. Abdominal ultrasonography revealed mild splenomegaly. Transthoracic echocardiography confirmed dextrocardia with structurally normal cardiac chambers and preserved cardiac function.

High-resolution contrast-enhanced computed tomography (CT) of the chest demonstrated situs inversus totalis with complete mirror-image transposition of the thoracic and abdominal viscera, including dextrocardia, a right-sided aortic arch, a left-sided liver, and right-sided spleen and stomach [Figure 2]. Extensive bilateral cystic bronchiectasis with diffuse bronchial wall thickening was identified. Multiple dilated bronchi contained air-fluid levels, consistent with retained infected secretions. Patchy bilateral lower lobe consolidations and numerous centrilobular tree-in-bud nodules involving the right upper and middle lobes indicated superimposed bronchopneumonia with endobronchial spread of infection. Marked bilateral bronchial artery enlargement was also observed. Mosaic attenuation involving both upper lobes was consistent with air trapping secondary to small airway disease [Figure 3].

Routine laboratory investigations were performed to assess systemic infection and baseline organ function [Table 1]. The complete blood count demonstrated mild anemia, while the total leukocyte count and differential were within normal limits. Inflammatory markers showed an elevated erythrocyte sedimentation rate (ESR) of 32 mm/h with a normal C-reactive protein (CRP). Liver function tests, renal

function tests, and blood glucose levels were all within normal limits. Sputum culture grew pan-susceptible *Klebsiella* spp., confirming a superimposed bacterial respiratory infection and supporting targeted antimicrobial treatment.

Taken together, the patient's lifelong history of recurrent sinopulmonary infections, chronic rhinosinusitis, recurrent otitis media, progressive bronchiectasis, and complete situs inversus totalis with dextrocardia raised a strong clinical suspicion of primary ciliary dyskinesia (PCD) with Kartagener syndrome. Several potential causes of chronic bronchiectasis and recurrent respiratory infections were considered. Post-tuberculosis bronchiectasis was an important differential diagnosis because the patient had previously received treatment for presumed pulmonary tuberculosis. However, according to the family, the diagnosis had been made by a local general practitioner in her rural community, and no records of microbiological testing, chest imaging, or other confirmatory investigations were available. The basis of the original tuberculosis diagnosis therefore could not be independently verified and appeared to have been primarily clinical. Moreover, the patient's respiratory symptoms had begun in early childhood and followed a lifelong pattern of recurrent lower respiratory infections, chronic rhinosinusitis, recurrent otitis media, and progressive bronchiectasis. This pattern, together with complete situs inversus totalis and dextrocardia, was considered more consistent with an underlying disorder of mucociliary clearance than with isolated post-tuberculosis lung disease.

Other recognized causes of childhood-onset bronchiectasis, including cystic fibrosis, primary immunodeficiency, and chronic aspiration, were also considered clinically. However, the combination of complete situs inversus totalis with chronic upper- and lower-airway disease strongly favored PCD with Kartagener syndrome. Cystic fibrosis-specific testing and comprehensive immunological evaluation were not available in our setting and therefore could not be performed. The history of intermittent coughing with swallowing also raised the possibility of aspiration as a contributing factor; however, this did not adequately explain the patient's longstanding sinonasal and otologic disease or the associated laterality abnormality. Thus, while alternative causes could not be definitively excluded because of limited diagnostic resources, the overall clinical and radiological phenotype was considered highly characteristic of Kartagener syndrome [Table 4].

Retrospective application of the PICADAR score further supported the clinical probability of PCD. The radiological findings demonstrated complete situs inversus totalis and extensive bilateral cystic bronchiectasis with associated air trapping and features of superimposed infection. Based on the combined clinical history, characteristic laterality abnormality, radiological findings, and supportive PICADAR assessment [Table 2], a clinical diagnosis of Kartagener syndrome was made. However, PCD-specific confirmatory investigations, including nasal nitric oxide measurement, ciliary motility analysis, high-speed video microscopy, transmission electron microscopy, immunofluorescence studies, and genetic testing, were unavailable in our setting and therefore could not be performed. Accordingly, the diagnosis should be regarded as a clinical diagnosis of Kartagener syndrome rather than genetically or physiologically confirmed PCD. A notable limitation of this report is that sputum acid-fast bacilli (AFB)

smear was not performed due to facility-level unavailability, and GeneXpert MTB/RIF molecular testing remains unavailable nationally.

In view of the acute infective exacerbation at presentation and the underlying structural lung disease, inpatient management was initiated promptly. The patient was admitted for five days and treated with intravenous ampicillin-sulbactam and clarithromycin to manage the acute respiratory infection. She also received inhaled bronchodilator therapy, airway-clearance measures, and supportive care throughout her hospitalization, resulting in gradual improvement in her respiratory symptoms.

A structured chest physiotherapy program was initiated to facilitate airway clearance because impaired mucociliary function in Kartagener syndrome leads to chronic retention of respiratory secretions. The regimen consisted of nebulized normal saline for 10–15 minutes, followed by postural drainage with chest percussion for 15–20 minutes, active cycle of breathing techniques (ACBT) or positive expiratory pressure (PEP) therapy for 10–15 minutes, huff coughing to facilitate sputum expectoration, and 20–30 minutes of aerobic exercise. These techniques were used to facilitate mucus clearance in the setting of impaired ciliary function.

Owing to her history of recurrent sinonasal disease and otologic symptoms, she subsequently underwent evaluation by an ENT specialist. Examination identified a small foreign body in the external auditory canal, which was removed, and the ear was cleaned. No other abnormalities were detected on otologic examination, and pure-tone audiometry demonstrated normal bilateral hearing. Further confirmatory investigations for primary ciliary dyskinesia, including ciliary motility analysis, were recommended; however, these specialized tests were unavailable in our resource-limited setting and could not be performed. Owing to the unavailability of confirmatory diagnostic modalities, including nasal nitric oxide measurement, ciliary functional or ultrastructural analysis, and genetic testing at our institution, referral to an international tertiary center in a neighboring country was recommended. However, the patient's family was unable to pursue overseas evaluation because of severe financial and socioeconomic constraints.

Upon discharge, the patient was prescribed oral azithromycin and daily guaifenesin syrup. She was discharged with instructions to continue daily chest physiotherapy at home. Given that the patient resided in a remote rural area with limited access to specialized respiratory rehabilitation services, considerable emphasis was placed on patient and family education before discharge. Detailed counseling was provided regarding the chronic nature of Kartagener syndrome, the importance of adherence to airway-clearance therapy, early recognition of respiratory exacerbations, and the need for regular follow-up. The patient and her caregivers received practical training in home-based chest physiotherapy techniques, including breathing exercises, postural drainage, percussion, and huff coughing. Demonstration videos and supervised practice sessions were used to ensure that the family could safely perform these techniques independently after returning to their home community.

Although the patient had not yet reached reproductive age, counseling regarding the potential long-term reproductive implications of PCD was considered relevant. The patient and her family were informed

about the potential for future subfertility and the increased risk of ectopic pregnancy associated with impaired tubal ciliary function. The family was therefore advised regarding preconception counseling, early antenatal assessment, and prompt medical evaluation of any future pregnancy.

Given that the patient resided in a remote rural village located hundreds of kilometers from our tertiary center, direct in-person follow-up posed significant logistical challenges. To ensure continuity of care, a collaborative follow-up plan was established with a local general practitioner at a primary care clinic in the patient's hometown. Additionally, longitudinal monitoring was facilitated via structured remote telemedicine consultations.

At the 1-month post-discharge follow-up, conducted through a structured telemedicine consultation with the patient and her family, substantial clinical improvement was reported. Her daily cough had markedly decreased, with a corresponding reduction in sputum production, and she reported no further episodes of hemoptysis. Dizziness and severe fatigue had resolved, and her overall activity level had increased. She was able to return to school and participate more actively in routine daily activities, with improved exercise tolerance compared with her pre-admission status. The patient and her mother reported continued adherence to the prescribed daily chest physiotherapy regimen, including the airway-clearance techniques demonstrated before discharge, and confirmed that they were able to perform the techniques correctly at home. Adherence to oral azithromycin was also maintained. No significant respiratory exacerbation was reported during the first month following discharge. Telemedicine follow-up was successfully coordinated with the patient and her family despite their residence in a remote rural area, allowing clinical progress and treatment adherence to be assessed without requiring immediate travel to the tertiary hospital. Follow-up beyond the first month was not available at the time of manuscript preparation. Continued clinical follow-up was planned at 3-month intervals to monitor respiratory symptoms, exacerbations, adherence to airway-clearance therapy, treatment tolerance, and functional status.

The patient's mother described a prolonged history of recurrent respiratory illnesses that had resulted in repeated visits to healthcare facilities and courses of treatment without a clear explanation for the underlying cause. The family reported that understanding the diagnosis and the chronic nature of the condition helped them better understand why the patient's respiratory symptoms had repeatedly returned despite previous treatment. Following counseling regarding Kartagener syndrome, the family gained a clearer understanding of the importance of regular airway clearance, adherence to prescribed medications, recognition of early respiratory deterioration, and timely communication with healthcare providers when symptoms worsened. The patient and her mother reported that the practical training in chest physiotherapy was particularly helpful because they were able to continue the techniques independently at home despite living far from specialized respiratory services.

The family perceived a meaningful improvement in the patient's breathing, physical activity, exercise tolerance, and ability to attend school following discharge. They also reported greater confidence in managing her condition at home and in recognizing symptoms that might indicate an exacerbation. The

family was advised to contact the treating team promptly if the patient developed worsening cough, increased sputum production, hemoptysis, fever, respiratory distress, or other concerning symptoms. The mother expressed that the explanation of the underlying condition and the practical guidance regarding long-term management were helpful in reducing uncertainty surrounding the patient's recurrent illness and improving their ability to participate in her care.

Table 2
PICADAR Score Calculation & Comparison

PICADAR Criteria	Standard Points	Patient Findings	Points Awarded
Full-term gestation	+ 2	Born at full term	2
Neonatal chest symptoms	+ 2	Not documented	0
Neonatal unit admission	+ 2	Not documented	0
Situs abnormality	+ 4	Complete situs inversus totalis confirmed on imaging	4
Perennial rhinitis	+ 1	Lifelong chronic rhinosinusitis and bilateral nasal polyps	1
Chronic ear/hearing problems	+ 1	Lifelong recurrent chronic otitis media	1
Congenital heart disease	+ 2	Echocardiogram normal (no structural cardiac defects)	0
Total PICADAR Score	14 max	—	8 / 14

PICADAR (Primary Ciliary Dyskinesia Rule) is a validated seven-item clinical prediction tool developed to estimate the likelihood of primary ciliary dyskinesia (PCD) in patients presenting with chronic respiratory symptoms, particularly persistent wet cough beginning in early childhood. The total score ranges from 0 to 14, with higher scores supporting an increased likelihood of Kartagener syndrome. In the present case, retrospective application of the PICADAR tool yielded a score of 8/14, indicating a high clinical probability of PCD and further supporting the clinical suspicion of Kartagener syndrome in the context of the patient's characteristic phenotype. This scoring assessment was particularly relevant given the absence of specialized diagnostic investigations, including ciliary functional studies and genetic testing, in our setting.

Table 1
Laboratory values

Lab Results	Patient Results	References
WBC	6.73	4.0–10.0 x1000/mm ³
Neutrophils (%)	54.1	50–70%
Hemoglobin (HGB)	11.5	11.7–18.0 g/dL
MCV	88.3	80–100 fL
MCH / MCHC	26.9 / 30.4	27–34 pg / 32–36 g/dL
Platelets	334	100–430 x1000/mm ³
Serum Creatinine	0.61	0.50–1.35 mg/dL
Blood Urea nitrogen	11	10–45 mg/dL
Sodium (Na+) / Potassium (K+)	138 / 3.9	135–150 mEq/L / 3.5–5.5 mEq/L
Chloride (Cl-)	109	95–106 mEq/L
AST / ALT / GGT	20 U/L / 8 U/L / 17 U/L	0–31 / 0–45 / 1–51
Total Bilirubin	0.24	0.30–1.35 mg/dL
Urinalysis (Leukocyte Esterase / Nitrite / Protein)	Negative	Negative
ESR	32	0–20 mm/h
CRP	6	0–10 mg/L
Procalcitonin	0.1	0–0.5 ng/mL

Hgb hemoglobin, WBC white blood cells, ALT alanine aminotransferase, AST aspartate aminotransferase, ALP alkaline phosphatase, ESR erythrocyte sedimentation rate, AST Aspartate Aminotransferase, ALT Alanine Aminotransferase, GGT Gamma-Glutamyl Transferase, CRP C-Reactive Protein

Table 3
Timeline of the patient's clinical course

Time	Clinical events
Early childhood	Recurrent productive cough and lower respiratory tract infections
Childhood	Recurrent sinusitis, chronic nasal obstruction and otitis media
2019	Nasal polypectomy
Subsequent years	Persistent respiratory symptoms and recurrent antibiotic-treated infections; previous treatment for pulmonary tuberculosis
Adolescence	Progressive exertional dyspnea, daily sputum production, weight loss and digital clubbing
Several months before admission	Intermittent hemoptysis and worsening exercise intolerance
2 weeks before admission	Marked worsening of productive cough, purulent sputum and hemoptysis
Current admission	Examination, laboratory investigations, CXR, echocardiography and CT performed
During hospitalization	Diagnosis of clinically suspected Kartagener syndrome; IV antibiotics and airway-clearance therapy initiated
Day 5	Clinical improvement and discharge
Post-discharge	Home chest physiotherapy, azithromycin and local/telemedicine follow-up
1 month	Reduced cough and sputum, resolution of respiratory distress, return to school

Table 3. Timeline of the patient's clinical course, from the onset of recurrent respiratory symptoms in early childhood through diagnosis, treatment, discharge, and one-month follow-up.

Table 4
Differential diagnoses considered in the evaluation of chronic bronchiectasis.

Differential diagnosis	Findings supporting consideration	Findings favoring Kartagener syndrome / against alternative
Kartagener syndrome / PCD	Lifelong wet/productive cough, recurrent lower respiratory infections, chronic rhinosinusitis, recurrent otitis media, extensive bronchiectasis, situs inversus totalis, dextrocardia	Characteristic combination of upper- and lower-airway disease with complete laterality abnormality strongly supports the diagnosis
Post-tuberculosis bronchiectasis	Post-tuberculous bronchiectasis typically manifests as localized, asymmetrical, upper-lobe fibro-cavitary scarring and traction bronchiectasis. In contrast, our patient demonstrated diffuse, bilateral cystic bronchiectasis with lower-lobe predominance.	Original TB diagnosis was clinically made without available microbiological or imaging documentation; Post-TB lung damage cannot account for the patient's lifelong history of upper airway disease dating back to early childhood (recurrent otitis media, chronic rhinosinusitis, recurrent nasal polyposis, and anosmia) nor her situs inversus totalis.
Cystic fibrosis	Chronic productive cough, recurrent respiratory infections, bronchiectasis	Complete situs inversus is not characteristic; no documented history of typical CF manifestations; CF-specific testing unavailable
Primary immunodeficiency	Recurrent respiratory infections from childhood	No known history of unusual/opportunistic infections or other manifestations of immunodeficiency; characteristic laterality abnormality strongly favors PCD; comprehensive immunologic testing unavailable
Other causes of non-CF bronchiectasis	Chronic structural lung disease and recurrent infections	The combination of chronic upper-airway disease, otitis media, bronchiectasis, and situs inversus strongly favors a congenital mucociliary disorder

Table 4. Differential diagnoses considered in the evaluation of childhood-onset recurrent respiratory infections and bronchiectasis, with clinical features supporting or favoring Kartagener syndrome.

Discussion

Primary ciliary dyskinesia (PCD) is a genetically heterogeneous autosomal recessive disorder caused by structural or functional abnormalities of motile cilia, resulting in impaired mucociliary clearance and chronic respiratory disease. Approximately 50% of individuals with PCD exhibit laterality abnormalities, most commonly situs inversus totalis; the coexistence of PCD and situs inversus is classically referred to as Kartagener syndrome [1–3]. The estimated incidence of PCD is approximately 1 in 30,000 to 1 in 40,000 live births, although the true frequency is likely underestimated because of delayed or missed diagnosis [1, 3, 8].

Our patient demonstrated the characteristic multisystem phenotype of Kartagener syndrome, with recurrent lower respiratory tract infections, chronic productive cough beginning in early childhood, chronic rhinosinusitis, recurrent otitis media, progressive bilateral bronchiectasis, and complete situs inversus totalis with dextrocardia. Pathogenic variants in genes encoding structural components of ciliary dynein arms, including *DNAH5* and *DNAI1*, are among the recognized genetic causes of PCD [2, 4]. A high prevalence of parental consanguinity among clinically unaffected parents of affected patients is frequently discussed in the reported literature, including cases by Ibrahim et al. and Gebray et al. [1, 2]. Consanguinity may increase the likelihood that both parents carry the same rare recessive pathogenic variant, supporting the possibility of an autosomal recessive disorder. In our patient, the presence of parental consanguinity was therefore consistent with an inherited recessive condition, although molecular testing was not available to identify the underlying genetic variant. This finding emphasizes the importance of considering inherited disorders in patients with characteristic phenotypes, particularly in populations in which consanguineous unions are common.

The clinical manifestations of PCD are heterogeneous and may emerge during the neonatal period or later in childhood. Persistent wet cough, recurrent lower respiratory tract infections, chronic rhinosinusitis, recurrent otitis media, and progressive bronchiectasis are characteristic respiratory manifestations [5–9]. Abnormal embryonic ciliary function may also disrupt left–right axis determination, resulting in dextrocardia or situs inversus totalis. PCD can additionally have important reproductive consequences, including subfertility and an increased risk of ectopic pregnancy in females resulting from impaired tubal ciliary motility required for ovum transport, as well as infertility related to abnormal sperm flagellar motility in males [6–9].

Some neonates may experience unexplained respiratory distress at birth, whereas older children typically develop a chronic wet, mucus-producing cough associated with impaired mucociliary clearance and evolving bronchiectasis, together with asthma-like symptoms, chronic rhinosinusitis, and recurrent otitis media. As the condition advances, clinical and radiographic signs of bronchiectasis may become evident [1, 2]. The coexistence of chronic upper- and lower-airway disease with complete situs inversus totalis in our patient was therefore strongly suggestive of Kartagener syndrome. This clinical presentation closely parallels the documented literature; for instance, Gebray et al. reported a similar clinical picture characterized by a long-standing productive cough and a history of multiple treatments for pneumonia [2]. Recognizing these overlapping features early in pediatric practice remains important, as PCD should be considered in children with recurrent disease involving both the upper and lower respiratory tracts, particularly when laterality abnormalities are also present.

An important diagnostic consideration in this patient was the reported history of previous tuberculosis treatment. In a setting where tuberculosis is common and diagnostic resources are limited, recurrent respiratory symptoms and established bronchiectasis may readily be attributed to previous or recurrent pulmonary tuberculosis. In this case, however, the original diagnosis of tuberculosis could not be independently verified because treatment had reportedly been initiated by a local general practitioner without available microbiological, radiological, or other confirmatory documentation. More importantly,

tuberculosis did not adequately account for the patient's lifelong pattern of recurrent sinopulmonary disease, chronic rhinosinusitis, recurrent otitis media, and complete situs inversus totalis. This illustrates an important diagnostic challenge in resource-limited settings: common infectious diagnoses may coexist with, or potentially obscure, an underlying congenital disorder of mucociliary clearance. Recognition of laterality abnormalities in a patient with longstanding bronchiectasis should therefore prompt consideration of PCD even when previous infectious diagnoses have been made.

Although definitive diagnostic testing is preferred for confirmation of PCD, such testing was not accessible in our setting. The diagnosis in our patient was therefore supported by the characteristic clinical phenotype, the presence of situs inversus totalis with dextrocardia, extensive bronchiectasis, and structured clinical assessment. In this context, the PICADAR score provided additional support for the clinical suspicion of PCD. However, the absence of molecular and specialized ciliary testing represents an important diagnostic limitation and should be acknowledged when interpreting the diagnosis. This highlights a broader challenge in resource-limited settings, where limited access to specialized diagnostic facilities may contribute to delayed recognition of PCD and allow progressive structural lung disease to develop before diagnosis.

As there are currently no curative treatment options, management of PCD is directed primarily toward preventing and treating disease complications. Treatment recommendations are largely based on expert opinion and extrapolation from cystic fibrosis and non-cystic fibrosis bronchiectasis guidelines, despite the distinct pathophysiology of PCD. Consequently, treatment approaches have varied considerably across different countries and clinical settings. Among the available therapeutic options, azithromycin maintenance therapy has demonstrated a beneficial effect on respiratory exacerbations and represents one of the better-supported pharmacological interventions in PCD [6–10, 12]. Tadesse et al. described clinical improvement following antibiotics, mucolytic therapy, and chest physiotherapy, followed by long-term prophylactic antimicrobial therapy [7]. Gebray et al. reported management with thoracic physiotherapy, mucolytic therapy, and intermittent azithromycin, with no exacerbations reported during follow-up [2]. Arunabha et al. used antibiotics, bronchodilator therapy, and mucolytic treatment with subsequent clinical improvement, whereas Ibrahim and Daood reported a severe pediatric presentation requiring ventilatory support for acute respiratory failure [1, 3]. Collectively, these reports highlight the central role of infection control and airway clearance while demonstrating that treatment intensity varies according to disease severity and the availability of follow-up resources [1–3]. In our patient, intravenous ampicillin-sulbactam and clarithromycin were administered during the acute presentation, together with inhaled bronchodilator therapy and structured airway-clearance interventions, followed by oral azithromycin and a mucolytic after discharge.

A retrospective study conducted in China included patients with PCD diagnosed between 2009 and 2021 and evaluated differences in clinical outcomes, pulmonary function, and chest CT findings between those who received azithromycin and those who did not. Patients treated with azithromycin experienced fewer respiratory exacerbations and demonstrated improved exercise tolerance compared with those who did not receive the medication. The potential benefit of azithromycin in PCD is thought to extend

beyond its antimicrobial activity, particularly through its anti-inflammatory effects, which may help reduce airway inflammation and mucus production. In selected patients with recurrent respiratory exacerbations, prolonged low-dose azithromycin may therefore be considered as part of individualized long-term management [2, 13]. However, its use should be considered within the context of the limited PCD-specific evidence and the potential for antimicrobial resistance and other adverse effects.

Bronchiectasis and recurrent respiratory infections in PCD are managed through a combination of airway-clearance techniques, appropriate antimicrobial therapy, and, when clinically indicated, bronchodilator and mucolytic therapy. Airway-clearance physiotherapy plays a central role in promoting secretion clearance and limiting the accumulation of mucus within structurally abnormal airways. Regular physical exercise may also support sputum clearance and improve functional capacity. Vaccination against influenza and pneumococcal disease is an important component of preventive respiratory care. Overall, effective respiratory management is essential to reduce recurrent infections and potentially limit further progression of structural lung disease following diagnosis [2, 3, 13].

Airway clearance represented the central component of long-term management in our patient. Before discharge, she received supervised training in postural drainage, chest percussion, breathing techniques, huff coughing, and other airway-clearance measures. Because she returned to a rural community without access to specialized respiratory rehabilitation services, particular emphasis was placed on family education and the development of a sustainable home-based physiotherapy routine. Demonstration videos and supervised practice were used to ensure that the patient and her mother could continue these techniques independently. This practical adaptation is particularly relevant in resource-limited settings, where continuity of specialist respiratory care may be difficult to maintain.

Managing chronic suppurative lung disease in patients from remote, rural areas requires practical adaptations to overcome geographic and socioeconomic barriers. Because our patient lived in rural town far from specialist care, routine in-person hospital visits were unfeasible. We addressed this limitation by coordinating care with a local community physician in her home district and using remote telemedicine consultations to monitor her recovery. At the 1-month post-discharge evaluation, this hybrid follow-up approach confirmed substantial symptomatic improvement, evidenced by a reduced cough burden and the patient's successful return to school. Continued follow-up is planned at 3-month intervals to monitor symptoms, respiratory infections, adherence to airway-clearance therapy, and overall clinical status. This experience highlights the potential role of decentralized primary-care coordination and digital health tools in maintaining long-term respiratory care for complex pediatric cases in resource-limited settings. Given the absence of a universally standardized treatment protocol and the limited disease-specific evidence base, further studies are needed to establish more consistent and evidence-based approaches to long-term management.

PCD may also have reproductive consequences because impaired ciliary motility can affect sperm transport and female tubal function. In female patients, this may contribute to subfertility and an increased risk of ectopic pregnancy. Although the patient was 15 years old and had not yet reached

reproductive age, reproductive counseling was considered appropriate because marriage and pregnancy may occur at a relatively young age in some rural communities in the patient's setting. The patient and her mother were therefore provided with age-appropriate education regarding the potential reproductive implications of PCD, including future subfertility and ectopic pregnancy risk, as well as the importance of preconception counseling, early antenatal care, and prompt medical evaluation during any future pregnancy. This counseling was intended to provide the family with appropriate information in advance rather than to address an immediate reproductive concern.

This case illustrates that Kartagener syndrome can remain unrecognized for years despite a classic clinical phenotype, particularly in resource-limited settings where recurrent respiratory infections may be managed episodically and specialized PCD testing is inaccessible. By adolescence, delayed recognition in this patient was associated with extensive bilateral cystic bronchiectasis and significant functional limitation. Nevertheless, recognition of the combination of lifelong wet cough, recurrent sinonasal and otologic disease, bronchiectasis, and situs inversus totalis allowed a clinically supported diagnosis to be made. The subsequent management strategy demonstrates that, even when specialized respiratory services are limited, sustained airway-clearance therapy can be adapted to the home environment through family education, supervised training, local primary-care coordination, and telemedicine. To our knowledge, this is the first reported case of Kartagener syndrome from Somalia and adds to the limited literature describing the recognition and practical management of PCD in resource-constrained settings.

This case has several strengths. First, the patient demonstrated a highly characteristic Kartagener phenotype, with lifelong recurrent sinopulmonary disease, chronic upper-airway and otologic involvement, extensive bronchiectasis, and complete situs inversus totalis. Second, the case illustrates the practical diagnostic challenges encountered in a resource-limited setting and demonstrates how clinical assessment and radiological findings can support recognition when specialized PCD investigations are unavailable. Third, the report describes a pragmatic management strategy based on family-centered airway-clearance training, home-based physiotherapy, local primary-care coordination, and telemedicine follow-up.

Several limitations should nevertheless be acknowledged. The diagnosis could not be confirmed using PCD-specific investigations, including nasal nitric oxide measurement, ciliary functional or ultrastructural analysis, immunofluorescence, or genetic testing, because these services were unavailable locally and international referral was not financially feasible. The PICADAR score provides supportive clinical evidence but is not a substitute for definitive diagnostic testing. In addition, the patient's previous diagnosis and treatment for tuberculosis could not be independently verified because the original clinical records, microbiological results, and imaging were unavailable. Comprehensive evaluation for other causes of bronchiectasis, including cystic fibrosis and primary immunodeficiency, was also limited by the diagnostic resources available. Finally, follow-up beyond one month was unavailable at the time of manuscript preparation, limiting assessment of long-term clinical response, exacerbation frequency, pulmonary function, and treatment tolerance.

Conclusion

Kartagener syndrome is a rare manifestation of primary ciliary dyskinesia that should be considered in patients presenting with recurrent sinopulmonary infections, chronic rhinosinusitis, bronchiectasis, and laterality abnormalities such as dextrocardia or situs inversus totalis. This case highlights the challenges of delayed diagnosis in resource-limited settings, where access to specialized diagnostic investigations may be limited. Nevertheless, careful clinical assessment combined with characteristic radiological findings can facilitate recognition of the disease and initiation of appropriate management when definitive diagnostic testing is unavailable. Early recognition, sustained airway-clearance therapy, appropriate treatment of respiratory infections, multidisciplinary follow-up, and comprehensive patient and family education are essential components of long-term care and may help limit further pulmonary deterioration and improve quality of life. To our knowledge, this is the first reported case of Kartagener syndrome from Somalia, highlighting both the geographic underrepresentation of the condition and the practical challenges of providing long-term care in resource-limited settings.

Abbreviations

KS: Kartagener syndrome; ACBT, Active Cycle of Breathing Techniques; ALT, Alanine Aminotransferase; AST, Aspartate Aminotransferase; CRP, C-Reactive Protein; CT, Computed Tomography; CXR, Chest X-Ray; ENT, Ear, Nose, and Throat; ESR, Erythrocyte Sedimentation Rate; FEV₁, Forced Expiratory Volume in One Second; FVC, Forced Vital Capacity; GGT, Gamma-Glutamyl Transferase; HGB, Hemoglobin; MCH, Mean Corpuscular Hemoglobin; MCHC, Mean Corpuscular Hemoglobin Concentration; MCV, Mean Corpuscular Volume; PCD, Primary Ciliary Dyskinesia; PEP, Positive Expiratory Pressure; PICADAR, Primary Ciliary Dyskinesia Rule; WBC, White Blood Cell Count.

Declarations

Ethics approval

Ethical approval was waived/not required for this single case report according to institutional policy. The patient was managed according to standard clinical practice.

Clinical trial number:

Not applicable.

Consent for publication

Written informed consent was obtained from the Patient's mother for the publication of this case report

Availability of data and materials

All relevant data related to this case are included in the manuscript. Additional information is available from the corresponding author upon reasonable request.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

All authors contributed to the clinical management of the patient, data collection, literature review, manuscript preparation, and final approval of the manuscript.

References

1. Ibrahim R, Daood H. Kartagener syndrome: A case report. *Can J Respir Ther*. 2021;57:44–8. <https://doi.org/10.29390/cjrt-2020-064>.
2. Gebray HM, Chekol AL, Mihiretie SA, et al. Kartagener syndrome in a young Ethiopian boy: a case report. *J Med Case Rep*. 2025;19:230. <https://doi.org/10.1186/s13256-025-05245-8>.
3. Arunabha DC, Sumit RT, Sourin B, Sabyasachi C, Subhasis M. Kartagener's syndrome: a classical case. *Ethiop J Health Sci*. 2014;24(4):363–8. <https://doi.org/10.4314/ejhs.v24i4.13>.
4. Afzelius BA. A human syndrome caused by immotile cilia. *Science*. 1976;193(4250):317–9. <https://doi.org/10.1126/science.1084576>.
5. Noone PG, Bali D, Carson JL, Sannuti A, Gipson CL, Ostrowski LE, Bromberg PA, Boucher RC, Knowles MR. Discordant organ laterality in monozygotic twins with primary ciliary dyskinesia. *Am J Med Genet*. 1999;82(2):155–60. [https://doi.org/10.1002/\(sici\)1096-8628\(19990115\)82:2%3C155::aid-ajmg11%3E3.0.co;2-t](https://doi.org/10.1002/(sici)1096-8628(19990115)82:2%3C155::aid-ajmg11%3E3.0.co;2-t).
6. 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(8):913–22. <https://doi.org/10.1164/rccm.201301-0059CI>.
7. Tadesse A, Alemu H, Silamsaw M, et al. Kartagener's syndrome: a case report. *J Med Case Rep*. 2018;12:5. <https://doi.org/10.1186/s13256-017-1538-2>.
8. Lobo LJ, Zariwala MA, Noone PG. Ciliary dyskinesia: primary ciliary dyskinesia in adults. *Eur Respir Mon*. 2011;52:130–49. <https://doi.org/10.1055/s-0035-1546748>.

9. Chilvers MA, Rutman A, O'Callaghan C. Ciliary beat pattern is associated with specific ultrastructural defects in primary ciliary dyskinesia. *J Allergy Clin Immunol.* 2003;112(3):518–24.
[https://doi.org/10.1016/s0091-6749\(03\)01799-8](https://doi.org/10.1016/s0091-6749(03)01799-8).
10. Fraser RS, Muller NL, Colman N, Pare PD. Bronchiectasis and other bronchial abnormalities. In: Fraser RS, Muller NL, Colman N, Pare PD, editors. *Diagnosis of diseases of the chest.* 4th ed. Philadelphia: W.B. Saunders Company; 1999. pp. 2281–3.
11. Behan L, Dimitrov BD, Kuehni CE, Hogg C, Carroll M, Evans HJ, Goutaki M, Harris A, Packham S, Walker WT, Lucas JS. PICADAR: a diagnostic predictive tool for primary ciliary dyskinesia. *Eur Respir J.* 2016;47(4):1103–12. doi:10.1183/13993003.01551-2015. Available from:
<https://doi.org/10.1183/13993003.01551-2015>
12. Paff TOH, Nielsen KG, Haarman EG. Current and future treatments in primary ciliary dyskinesia. *Int J Mol Sci.* 2021;22(18):9834. <https://doi.org/10.3390/ijms22189834>.
13. Guan YZX, Yang H, Xu H, Zhao S. Long-term azithromycin treatment in pediatric primary ciliary dyskinesia: a retrospective study. *Front Pediatr.* 2022;10(10):905253.
<https://doi.org/10.3389/fped.2022>.

Figures



Figure 1

Chest radiography (posterior-anterior view) demonstrated dextrocardia with bilateral bronchiectatic changes along both the right and left cardiac borders

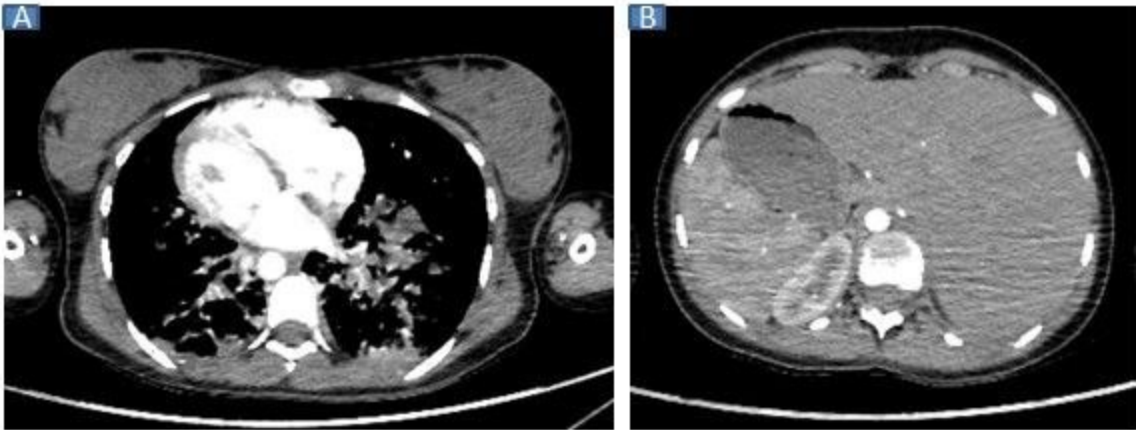


Figure 2

(A, B): Contrast-enhanced CT demonstrates situs inversus totalis with dextrocardia, a right-sided aortic arch, a left-sided liver, and right-sided spleen and stomach

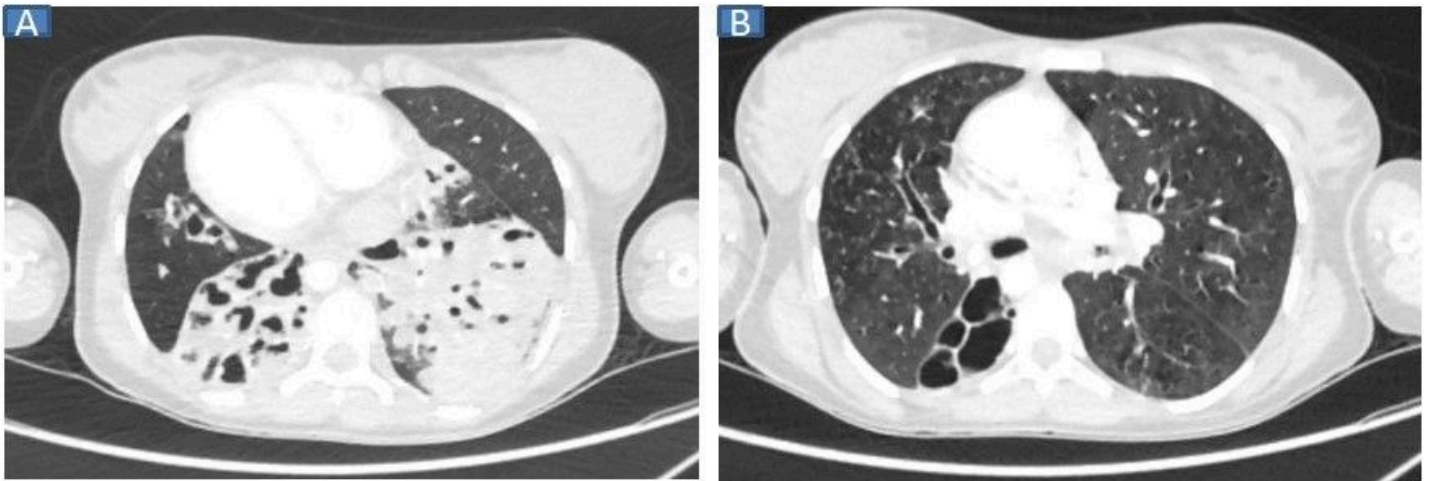


Figure 3

(A, B): Extensive bilateral cystic bronchiectasis with bronchial wall thickening, air-fluid levels, bilateral lower lobe consolidations, right upper and middle lobe tree-in-bud nodules, and upper lobe mosaic attenuation.