Kartagener's Triad
Review of the Literature and Report of a Case*

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In 1904, Siewert¹ reported a case of situs inversus with bronchiectasis in a 21-year-old white male. Guenther,² in 1923, described situs inversus and bronchiectasis in a 19-year-old girl who had two normal siblings, but a third sibling had situs inversus and "bronchial catarrh." Subsequently, in 1933, Kartagener³ added 11 cases of situs inversus and bronchiectasis, all of which also presented findings of chronic sinusitis. The association of these three abnormalities has since become known as Kartagener's Triad. Approximately 40 cases have been described in the literature to date.¹-¹³

The following case is presented because this patient is older than any heretofore reported, and because he exhibited a host of other pathological conditions, some of which may be related to the aforementioned group of abnormalities.

Case Report

A 63-year-old white male veteran of World War I was admitted to the hospital complaining of recurrent episodes of pain, redness, and swelling of his joints of 10 months' duration. He had had intermittent involvement of both knees, shoulders, elbows, wrists and of the small joints of both hands.

Past history revealed a chronic cough as far back as he could recall, unusual susceptibility to colds during childhood, and left otitis media at age 18. In 1918, he had acute sinusitis which was treated over a seven-month period with intranasal instillations of argyrol. Shortly thereafter, he developed a bluish discoloration of the skin. Following this acute episode of sinusitis, the cough became productive of about one cupful of purulent sputum daily. He claimed to have had numerous episodes of pneumonia during the past 30 years. His mother had had diabetes and one brother died of diabetes at age 13.

Physical examination revealed a well-developed, moderately obese, left-handed white male, who did not appear ill. Blood pressure 140/96, pulse 78, temperature 98.4 degrees Fahrenheit. There was bluish discoloration

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of the skin of the face, hands and scalp and purplish brown discoloration of the bulbar and palpebral conjunctiva. Bilateral ptterygia were present. A firm, non-tender mass, 1 centimeter in diameter, was noted in the buccal mucosa opposite the lower right first molar (subsequently excised and found to be sub-epithelial fibrosis). The nasal mucosa was edematous, and thick mucopurulent discharge was noted on the nasal floor. There was dullness to percussion and diminished breath sounds over the right lung base posteriorly. Loud, high-pitched rhonchi were heard throughout the remainder of the lung fields. The left diaphragm was

![Figure 1](image1.jpg)

**Figure 1**: Transposition of heart and aorta. Atelectasis of basilar segments of right lower lobe.—**Figure 2**: Atelectasis of basilar segments of right lower lobe visualized behind the heart.

![Figure 2](image2.jpg)

![Figure 3](image3.jpg)

**Figure 3**: Chest film made two years earlier than Figure 1. Large dilated air-containing bronchi can be seen in the area of pneumonia consolidation of the right lower lung field.—**Figure 4**: Bronchogram showing cylindrical and sac- cular dilatation of basilar bronchi.
higher than the right, but both moved adequately with respiration. The heart was percussed in the right hemithorax and was apparently of normal size. Liver dullness was percussed on the left. There was fusiform swelling of the proximal interphalangeal joints of the index and middle fingers of both hands. There was some pain on motion of all the joints of the upper limbs and of the large joints of the lower extremities. The prostate was moderately symmetrically enlarged and tender.

Complete blood count and urinalysis were normal. Sedimentation rate (Wintrobe, corrected) was 38 millimeters per hour. Sputa were negative for tubercle bacilli and positive for Streptococcus viridans, Neisseria catarrhalis and Friedlander's bacillus. Standard electrocardiogram revealed typical "mirror image" tracing of dextrocardia.

Chest fluoroscopy revealed the cardiac shadow in the right hemithorax. The aorta was also on the right side. A triangular shaped density in the right lower lung field was thought to represent atelectasis of the right lower lobe (Figures 1 and 2). Bronchograms made elsewhere two years previously revealed extensive saccular and cylindrical bronchiectasis in the basilar segments of both lower lobes (Figures 3 and 4). Sinus films revealed cloudiness of all sinuses except the sphenoids (Figure 5). Gastrointestinal studies revealed complete transposition of the abdominal viscera (Figures 6 and 7). Films of all large joints and hands did not show

**FIGURE 5:** Clouding of frontal sinuses with sclerosis and blurring of the sinus margins.
conclusive radiographic evidence of rheumatoid arthritis.

At bronchoscopy, the larynx and trachea appeared normal. On the right, only upper and lower lobe bronchi could be identified. Three divisions of the left main stem bronchus were seen. The bronchial mucosa was reddened, edematous and bled easily. Profuse mucopurulent secretion was present in both bronchial trees, most abundant in the right lower lobe bronchus.

Surgical treatment was not considered advisable because of the extensive involvement and because of the patient's age. He was, therefore, treated medically with penicillin inhalations and streptomycin intramuscularly. Marked relief of cough followed and sputum was reduced from 240 cubic centimeters to 30 cubic centimeters daily.

Salicylates and physiotherapy gave partial relief of joint symptoms.

Discussion

Siewert, in his original paper, speculated on the possible congenital etiology of bronchiectasis because of his observance of this condition in a patient who had congenital transposition of viscera. Proponents of the congenital origin of bronchiectasis have since enlarged upon this interesting observation, and subsequent reports of additional cases have been used to lend further support to this theory.

Adams and Churchill added to the literature five cases ranging from nine to 28 years of age. They observed that in 23 cases of situs inversus, five (21.7 per cent) had bronchiectasis; whereas, only 0.306 per cent of approximately 240,000 general hospital ad-

FIGURE 6  
Figure 6: Transposition of upper GI tract.

FIGURE 7  
Figure 7: Transposition of colon.
missions had bronchiectasis. According to Mallory,\textsuperscript{5} this high incidence of bronchiectasis in situ inversus may be accepted as indirect evidence in support of its congenital origin. Ingraham\textsuperscript{6} reported a case of Kartagener's Triad in whom multiple bilateral lobectomies were performed. The tissue was examined by the pathologist, who stated, "The entire picture strongly suggests a congenital malformation with persistence of peribronchial connective tissue and underdevelopment of alveolar structures." The observance of ectatic bronchi in infants shortly after birth may be accepted in support of the congenital theory. The occurrence of this disease in twins may also be used to strengthen this hypothesis.\textsuperscript{7}

Richards\textsuperscript{8} reports the case of a child with situs inversus, who from the age of five days was treated for nasal obstruction and cough, and who at the age of seven years showed x-ray evidence of sinusitis and bronchiectasis.

In a discussion of the genetics of the transposition of viscera, Cockayne\textsuperscript{9} presents evidence which he accepts as proof of the hereditary nature of this condition, and states that it is due to a rare recessive character. He claims that all organs are originally bilaterally symmetrical and change to spiral symmetry in the process of dextrorotation. In cases of situs inversus, the rotation is sinistral. He feels that, because malformations of the heart and bronchiectasis are more common in transposition of the viscera, the gene which produces sinistral rotation carries with it a greater liability to early death than does its dominant allelomorph for normal rotation.

Whether or not the evidence cited above is significant statistically to support the theory of congenital origin of bronchiectasis is debatable. Rosenthal,\textsuperscript{10} after his review of the literature, feels that there is insufficient evidence to support the assumption that a direct relationship exists between the anomaly of visceral transposition and bronchiectasis.

Unfortunately, a study of cases of Kartagener's Triad does not resolve the question as to whether the basic anomaly in congenital bronchiectasis is abnormal dilatation or abnormal structure of the bronchial wall present at birth and more easily attacked by recurring respiratory infections.

Practical consideration must be given to the observed high incidence of bronchiectasis in patients with situs inversus. This would include the use of all available prophylactic measures to prevent recurrent respiratory tract infections. In children with transposition of the viscera and a history of respiratory tract infections, early bronchography is recommended so that if bronchiectasis is present therapeutic measures may be instituted at once.
SUMMARY

1) A case exhibiting the typical features of Kartagener's Triad is reported.
2) The literature is reviewed and some of the bases of the congenital origin of bronchiectasis are discussed.
3) Attention is called to the need for early and vigorous efforts to prevent recurrent respiratory tract infections in patients with situs inversus.

RESUMEN

1) Se informa sobre un caso que exhibió los rasgos característicos del Terno de Kartagener.
2) Se repasa la literatura y se discuten algunas de las bases de la teoría del origen congénito de la bronquiectasia.
3) Se llama la atención sobre la necesidad de hacer esfuerzos tempranos y vigorosos para evitar repetidas infecciones del aparato respiratorio en pacientes con situs inversus.

REFERENCES

1 Siewert, A. K.: "Ueber einen Fall von Bronchiectasie bei einem Patien-


1904.
2 Guenther: Quoted by Adams and Churchill.
3 Kartagener: Quoted by Adams and Churchill.
4 Adams, R. and Churchill, E. D.: "Situs Inversus, Sinusitis, Bronchi-


6 Ingraham, R.: "A Case of Situs Inversus with Extensive Bilateral

Bronchiectasis Dating from Early Childhood and with Bilateral Lobec-


47:435, 1943.
8 Richards, W. F.: "Situs Inversus Viscerum, Absent Frontal Sinuses

with Ethmoid and Maxillary Infection, and Bronchiectasis. Kart-

gener's Triad," Tubercle, XXV:27, 1944.
9 Cockayne, E. A.: "The Genetics of Transposition of the Viscera,

10 Rosenthal, D. B.: "Bronchiectasis and Visceral Transposition with


12 Delp, M. H.: "Kartagener's Triad. Situs Inversus, Absent Frontal

Sinuses with Maxillary Ethmoid and Sphenoid Infection and Bron-

13 Cole, D. B. and Nalls, W. L.: "Situs Inversus, Sinusitis, and Bronchi-