Kartagener's Syndrome in Children

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The triad of situs inversus, bronchiectasis and sinusitis has borne the name of Kartagener since this author in 19331 collected 11 cases. The first case was reported by Siewert in 1904,2 in a 21 year old white male, the symptoms having been present since infancy. The literature has been recently well reviewed by Bergstrom, Cook, Scannell, and Berenberg.3 They collected 80 cases, including two of their own in a family where two additional siblings suffered from bronchiectasis and sinusitis without dextrocardia, and the father from sinusitis only. Zuckerman and Wurtzebach in 19514 accepted 40 cases which they collected from the literature as fulfilling the clinical requirements for acceptance to this disease. Their own case was in a 63 year old white male, with symptoms dating back to childhood.

In Bergstrom's and his colleagues' series only 16 of the 80 collected cases showed undisputable evidence of bronchiectasis roentgenologically, but in the larger percentage of those in whom history was available, the symptoms dated back to infancy or early childhood, and in 90 per cent symptoms were present before the age of 14 years. Richards5 reported a case in which symptoms were present on the third day of life.

Many cases described in the recent literature have been of dextrocardia without complete situs inversus, and as these have been generally accepted as valid cases, it would now probably be more fit to describe the syndrome as consisting of dextrocardia, bronchiectasis and sinusitis.

Following are the reports of five new cases.

Case 1: C.M., girl, white, age six weeks. This patient was first seen in Well Baby clinic, with no complaint. The birth weight was 6 pounds, 1 ounce, and she was now 7 pounds, 12 ounces. Physical examination revealed the heart to be on the right side with a loud systolic murmur heard best at the right of the sternum from the fourth to the sixth interspace. Over the chest there were rales with noisy inspiration and expiration. The liver edge could be felt on the left side. There was a slight cyanotic tinge to the skin, but more marked over the extremities. Erythrocyte count was 4,500,000 and hemoglobin 12.2 grams per cent. Roentgenograms showed complete transposition of all the viscera with marked cardiac enlargement. There was a markedly clear periphery of the lung parenchyma (Figure 1). There was little change in the patient's condition, except for frequent colds, until the age of four months, when she was hospitalized with acute pneumonia. From this time on progress was slow. At the age of seven months her weight was 11 pounds. She became increasingly cyanotic and dyspneic. Her erythrocyte count was now 7,800,000, with hemoglobin 11.9 grams per cent; marked microcytosis, polkilocytosis and polychromasia was present. Roentgenograms now showed increased cardiac enlargement, atelectasis of the right lower lobe, and marked pulmonary vascular engorgement of the left lung field (Figure 2). At the age of 10 months she had developed constant nasal discharge, yellow, frothy and mucoid,
Figure 3A. Case 1: Bronchogram at age of 10 months.
Figure 3B. Case 1: Spot film showing compression of right main-stem bronchus by soft tissue density.
both anteriorly and posteriorly, and roentgenograms revealed very small maxillary
sinuses, with no others visible. Bronchoscopy at this time showed the right main
bronchus compressed anteriorly and this side could not be entered further. Mucus
was aspirated from both sides. Bronchograms showed posterior displacement and
partial obstruction to the right main bronchus by soft tissue density, atelectasis
of the greater portion of the right lung with irregular small bronchial outlines
which could not be evaluated, and probably normal left bronchi (Figure 3). The
infant's condition remained the same for some months. She had many upper
respiratory infections, cyanosis and dyspnea continued, and she developed marked
clubbing of the distal phalanges of all digits. At the age of one year she weighed
13 pounds, 8 ounces. At 14 months she had another acute respiratory infection
and died 30 minutes after arrival at the hospital. At necropsy, there was dextro-
cardia with situs transversus. The heart was markedly distorted, and complete
transposition of the great vessels had occurred, i.e., the aorta on the left and
cardiac arteries on the right. Interventricular septal defect was also present.
Both lungs were reversed, with two lobes on the right, and four on the left. The
right lung was completely collapsed, meaty red and firm, and the left large and
fully expanded. The right main bronchus was completely collapsed, and the
cartilage in the posterior wall appeared to be absent. Microscopically the right
lung showed small focal hemorrhages, the adjoining parenchyma areas of emphy-
sema. There was diffuse disruption of many elastic fibrils. There were larger
areas of alveolar hemorrhage in the right lower lobe, infiltrated with numbers of
polymorphonuclears and macrophages. The left lower lobe had vast areas of
collapsed alveoli with their walls thickened, avascular, and appearing cellular.
Many bronchi were collapsed and others contained debris. The right main bron-
chus presented marked thinning with reduced staining reaction of the cartilage
plates. At this point there was angulation and compression of the bronchus wall
with narrowing or collapse of the lumen. The mucous membranes were thickened,
the glands prominent, and there was diffuse infiltration with lymphocytes and
plasma cells. The sinuses were not examined.

Case 2: P.R., girl, white, aged nine years. The complaint was repeated respira-
tory infections since birth. Atelectasis of the right lung was diagnosed at the age
of three days. There was frequent hospitalization during infancy and early child-
hood for fever, cough and difficult breathing, with cyanosis during the later
attacks. At 22 months she had purulent sputum. Pneumonia occurred at least
once yearly. There was a constantly running nose between the acute attacks of

![Figure 1](image1.png)

**Figure 1, Case 1:** Age six weeks. Complete transposition of viscera, cardiac enlargement, and clear lung parenchyma.

![Figure 2](image2.png)

**Figure 2, Case 1:** Age seven months. Atelectasis of right lower lobe.
cough and fever. Adenotonsillectomy was performed at five years. Physical examination showed a thin, underweight girl, with cough and moderate wheezing. There was mucous discharge from the nose and granular, slightly injected post-pharynx with purulent discharge. There were fine and course moist rales, inspiratory and expiratory, throughout the chest. Dextrocardia was present. There was slight clubbing of the fingers. Blood and urine were normal. Trypsin was repeatedly found to be present in the stool. Tuberculin and coccidioidin skin tests were negative. Roentgenograms showed ethmoid and maxillary sinusitis, dextrocardia without complete situs inversus, generalized pulmonary fibrosis, and probable bronchiectasis of the middle and lower left lobes. Bronchograms (Figure 4) showed atelectasis of the left middle lobe with severe saccular bronchiectasis, minimal cylindrical bronchiectasis of the left lower lobe, and possibly saccular bronchiectasis of the right lingula. Resection of the left middle lobe was performed with a wedge resection of the lower segment of the left upper lobe. Gross section of both pieces of lung showed large cylindrical thick-walled fibrotic bronchi filled with purulent material, and surrounded by small amounts of collapsed hyperemic lung tissue. Microscopic examination showed muscular and cartilagenous tissue absent in most of the peribronchial regions and numerous irregular alveolar spaces in the surrounding inflammatory tissue were lined by a conspicuous low cuboidal epithelium. There was dense infiltration of the bronchial walls and surrounding tissue with lymphocytes, plasma cells and eosinophiles in a loose vascular and fibrous stroma. Between the altered bronchi there were varying amounts of pulmonary tissue, some portions of which showed thin-walled distended alveoli, and some alveoli which were compressed, irregular, and contained macrophages, erythrocytes and lymphocytes. Convalescence was uneventful, and she was discharged considerably improved.

Figure 4, Case 2: Atelectasis of left middle lobe with saccular bronchiectasis.
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Case 3: R.T., girl, white, aged 10 years. The complaint was running nose, cough and wheezing since birth. For the past year these symptoms had increased in severity, and she had an afternoon fever. Her brother suffered from a similar condition (Case 4). Physical examination showed a thin, underweight girl. The tympanic membranes were retracted. There was muco-pus on the turbinates and in the post-pharynx. Tonsils were present, and palpable, shotty anterior cervical lymph-nodes. The heart was on the right side. The blood and urine were normal. Tuberculin and coccidioidin skin tests were negative. Roentgenograms showed complete situs inversus and pansinusitis; laminagram showed atelectasis of the left middle lobe (Figure 5). Bronchograms showed marked saccular bronchiectasis of the left middle lobe (Figure 6). Lobectomy was performed, and the gross specimen (Figure 7) showed numerous dilated bronchioles, with thick walls and diameters up to 4.5 mm. throughout the lobe. Histological examination showed most of the lung parenchyma collapsed. There was lymphocytic infiltration diffusely throughout the parenchyma, and in the lumen of the bronchioles some collections of polymorphonuclear leukocytes and lymphocytes in fibrinous material. There was rather marked diminution and destruction of elastic tissue and smooth muscle in the bronchial wall. Some of the bronchial cartilages were partially destroyed. The patient had an uneventful convalescence with marked improvement of her chest symptoms, with absence of fever and diminution of cough. However her sinusitis did not seem to have improved, either symptomatically or roentgenographically.

Figure 5, Case 3: Laminagram showing atelectasis of left middle lobe.
Case 4: C.T., boy, white, aged 14 years. The complaint was constantly running nose, chronic cough with sputum, and intermittent fever since early infancy. He had what was diagnosed as an asthmatic attack at five months, and had had them recurrently ever since. He had been treated for chronic sinusitis all his life and had a series of injections for "allergy" without benefit. Adenotonsillectomy was performed at nine years. He had been known to have had nasal polyps for years, with two operations for their removal at ages 10 and 12 years. His cough at entry was productive to the extent of one-half cup of yellowish-white, foul smelling sputum daily. He usually had a low grade fever in the evening. He had been hard of hearing for years. His sister (Case 3) suffered from similar complaints. Physical examination showed a tall thin boy with nasal speech. Both tympanic membranes were fibrotic and somewhat retracted. The right inferior turbinate was markedly hypertrophied, the middle had been removed. The left turbinates were swollen. There was much polypoid tissue on both sides with muco-pus. The posterior pharyngeal wall was covered with hypertrophied lymphoid tissue. In the

Figure 6, Case 3: Bronchogram showing saccular bronchiectasis of left middle lobe.
Figure 7, Case 3: Left middle lobe with dilated bronchioles.

Figure 8A, Case 4: Bronchogram showing tubular and slight saccular bronchiectasis of left middle lobe.
Figure 8B, Case 4: Lateral view.
chest there were coarse wheezes and groans bilaterally with both inspiration and expiration, and fine rales at both bases. Dextrocardia was present. White blood count was 21,100 with 81 per cent polymorphonuclears. Urine was normal. Tuberculin skin test was negative, coccidioidin positive. Roentgenograms showed evidence of pansinusitis, complete situs inversus, and chronic middle lobe disease of the left lung (atelectasis). Bronchograms revealed tubular and slight saccular bronchiectasis of the left middle lobe (Figure 8). Bilateral antral aspiration was done and pus obtained, new ostia were opened and flaps of nasal mucosa laid down. A polyp was removed on the right and a tonsillar tag from the left fossa. Two weeks later left middle lobe resection was performed. Microscopically there was widespread alteration of pulmonary architecture. Varying degrees of alveolar collapse were present with diffuse thickening of the walls of the alveoli by collagenous fibroelastic tissue. Much of the latter was infiltrated with plasma cells and lymphocytes. There were many dilated irregular bronchi and bronchioles, and these showed a replacement of their walls by a vascular fibroelastic tissue diffusely infiltrated with plasma cells, lymphocytes, and scattered eosinophiles. There were areas in the altered bronchioles filled with a polymorphonuclear exudate, and the surrounding parenchyma contained an occasional small abscess. Many irregular alveoli were lined with a dark cuboidal epithelium, and thick-walled arteries and arterioles were numerous. There were scattered areas of hemorrhage into alveoli. After an interval of three months the boy was much improved, his sputum had almost disappeared, but he still had some cough.

Case 5: P.B., girl, white, aged four years. The child came to the out-patient clinic because of pyoderma of the scalp. A history was elicited of chronic nasal discharge since early infancy. A cough was frequently present and this was always worse when she had a cold. She had some improvement after an adenotonsillectomy at two and a half years of age. Dextrocardia was known to have been present at the age of two. She had never had dyspnea, cyanosis, or fatigueability, and had always been very active. Her father had suffered from nasal allergies and sinus trouble. Physical examination showed a well nourished, apparently healthy girl. The nasal passages were filled with mucus which was also present on the post-pharyngeal wall. The heart was on the right. A sharp, high pitched, systolic murmur was heard at the aortic area and was transmitted down the right sternal border to the apex. Over the left middle chest there were inspiratory rales. The liver was palpable on the left. The blood and urine were normal. Tuberculin and coccidioidin skin tests were negative. Roentgenograms showed mucosal thickening or exudate in both antra and in the posterior ethmoid air cells, a complete situs inversus, and density in the left middle lobe area suggesting atelectasis or bronchiectasis. Bronchograms visualized all of the principal left bronchial branches and showed bronchiectasis of the left middle lobe with all of the rest of the lung appearing normal. Left middle lobe resection has been advised, but has not yet been performed.

Discussion

In the five cases of dextrocardia, four had complete situs inversus, one dextrocardia only. The case coming to necropsy was found to have other anomalies of the heart. All of the children had pulmonary atelectasis, one on the same side as the heart, the other four on the left side. Four had bronchiectasis in the collapsed lobes on the left, one (the youngest) beginning bronchiectasis or prebronchiectatic changes on the right. All five children had clinical evidence of sinusitis, four had roentgenologic evidence, and in one, the infant, the sinuses were undeveloped. Four of the cases were in girls, one in a boy. The ages at which the disease was diagnosed were 6 weeks, and 4, 9, 10 and 14 years. The 10 and 14 year old
patients were sister and brother. All three children having lobar resections showed considerable improvement of their pulmonary symptoms.

Bronchiectasis seems to accompany dextrocardia or complete visceral transposition in a large percentage of cases. Olsen\(^6\) reviewed the cases of 85 patients with dextrocardia, and found 14 (16.5 per cent) to have bronchiectasis, as contrasted with an incidence of less than 0.5 per cent among all patients at the Mayo Clinic. His study, and that by Adams and Churchill,\(^7\) lend support to the theory that the bronchiectasis in Kartagener's syndrome is congenital in origin. The theory is further supported by many cases in the literature in which the triad occurred in members of the same family, as reported by Bergstrom, et al.,\(^3\) Kaye and Meyer,\(^6\) and ourselves.

European authorities have long favored the congenital theory of origin of bronchiectasis, as opposed to the prevailing opinion in America that most bronchiectasis is acquired.\(^8\) That the congenital theory is not tenable in all cases is proved by the occurrence of the disease following the primary infection with tuberculosis, and following atelectasis from other causes. While in Kartagener's syndrome heredity must usually be the primary factor, or at least predisposing cause, it is conceivable, even probable, that pulmonary collapse may occur after birth in a lobe previously normal, and that bronchiectasis may follow. In the published reports on Kartagener's syndrome bronchiectasis is usually noted as being of the tubular or "varicose" type, instead of the cystic variety generally regarded as congenital.\(^9\) Kaye and Meyer reported a case in which the collapse and bronchiectasis may have been due to the presence of an anomalous left subclavian artery. In the first case of our series there was present a transposition of the great vessels, and the pulmonary changes were those of collapse with only beginning bronchiectasis or a pre-bronchiectatic condition of the walls. In three of our own cases the microscopic changes were degenerative rather than congenital. Bergstrom, et al., from the microscopic findings reported in their case that "the picture was in keeping with the acquired type of bronchiectasis."

Conway\(^9\) believes that the great majority of bronchiectases are acquired, and states: "If it is sometimes congenital it should be possible occasionally to demonstrate it in a stillborn foetus or in a neonate in whom atelectasis or infection have not occurred."

It is our own belief, influenced by a study of the literature and of our own cases, that the sequence of events in the development of Kartagener's syndrome is: 1) congenital anomaly of the cardiovascular system (dextrocardia, sometimes with other anomalies); 2) atelectasis; 3) bronchiectasis, and 4) sinusitis. That the early development of bronchiectasis is influenced by a developmental error in the bronchi themselves is possible, but direct proof is still lacking. There is considerable circumstantial evidence that a congenital factor of some sort is of importance.

The treatment of this disease consists of resection of the affected pulmonary tissue as soon as the patient is deemed a good surgical risk. The
age of the patient in the younger years, after the neonatal period, need not be considered in making the decision. Treatment of sinusitis is indicated, although it is usually not nearly so successful as that of the bronchiectasis.

SUMMARY

1) Five cases of Kartagener's syndrome are reported, all of whom had dextrocardia, atelectasis, early or late bronchiectatic changes, and sinusitis.

2) Lobectomy was performed on three with beneficial results. The youngest patient died before the operation could be performed.

3) From the previously reported cases, and from the present series, it may be concluded that a congenital factor of some sort plays a part in the pulmonary features of the disease. Whether there is a developmental error in the bronchial walls or whether the pulmonary lesions are secondary to the cardiovascular anomalies is still open to debate. In any case, it is probable that in most instances atelectasis precedes bronchiectasis.

RESUMEN

1) Se refieren cinco casos de síndrome de Kartagener, todos los cuales tenían dextrocardia, atelectasía, alteraciones bronquiectásicas tempranas o tardías y sinusitis.

2) En tres de ellos se realizó la lobectomía con resultados benéficos. El más pequeño de los enfermos murió antes de que la operación pudiese ser llevada a cabo.

3) De acuerdo con los casos antes relatados y con la serie presente, puede concluirse que hay un papel que desempeña un factor congénito en las características pulmonares de la enfermedad. Aun esta abierta la discusión acerca de si hay una falla en el desarrollo de las paredes bronquiales o si las lesiones pulmonares son secundarias a las anomalías cardiovascular. De cualquier modo, es probable que en la mayoría de los casos la atelectasía preceda a la bronquiectasía.

RESUME

1) L'auteur rapporte cinq observations de syndrome de Kartagener chez des enfants, dont tous étaient atteints de dextrocardie, d'atélectasie, de modifications bronchiectasiques précoces ou tardives, et de sinusite.

2) Une lobectomie fut pratiquée sur trois d'entre eux avec succès. Le plus jeune malade mourut avant que l'opération ait pu être pratiquée.

3) D'après les observations rapportées précédemment et d'après celle-ci, on peut conclure qu'un facteur congénital quelconque joue un rôle dans les symptômes pulmonaires de l'affection. Le débat reste ouvert au sujet de la question de savoir s'il s'agit d'une anomalie de développement des parois bronchiques, ou de lésions pulmonaires secondaires aux altérations cardio-vasculaires. En tout cas, il est probable que dans la plupart des observations, l'atélectasie précédé la bronchiectasie.
REFERENCES