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Dextrocardia and Corrected Transposition of the Great Arteries (I,D,D) in a Case of Kartagener's Syndrome: A Unique Association

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Summary: Kartagener's syndrome (KS) usually includes mirror-image dextrocardia. The incidence of congenital heart disease in KS is comparable with that in the general population. This paper reports on a case of Kartagener's syndrome associated with dextrocardia, corrected transposition of the great arteries (I,D,D), ventricular septal defect, and valvar pulmonary stenosis in an 8-year-old girl.

Key words: Kartagener's syndrome, corrected transposition of the great arteries, situs inversus

Introduction

Kartagener's syndrome (KS) is a triad of situs inversus, chronic sinusitis, and bronchiectasis. It was first described by Siewert in 1904, then established by Kartagener in 1933.[1,2] It was Afzelius in 1976 who, while studying sperm from infertile males with Kartagener's syndrome, determined that the disorder is caused by decreased ciliary motility and defined the ultrastructural abnormalities that characterize it.[1,2] Kartagener's syndrome constitutes around 50% of cases of the immotile cilia syndrome. It includes mirror-image dextrocardia. No other congenital malformations of the heart or other organs are apparent.

Corrected transposition of the great arteries is an unusual cardiac malformation, in which the normal hemodynamic pathways are not altered by the malformation. We report here a unique case of Kartagener's syndrome and dextrocardia, corrected transposition of the great arteries (I,D,D), ventricular septal defect, and valvar pulmonary stenosis in an 8-year-old girl.

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Case Report

The patient, an 8-year-old girl, born to first-degree consanguineous parents, presented to our hospital with a history of recurrent respiratory tract infections since infancy. These infections required frequent hospitalizations and antibiotic therapy. She had a history of snoring, mouth breathing, postnasal drip, chronic cough with purulent expectoration, and easy fatiguability.

On physical examination she was thin and pale, and in moderate respiratory distress. She had a right nasal polyp. Her lung exam revealed diffuse wheezes and rales, as well as marked decrease in air entry over the right lower lung field. She had right-sided heart sounds and a holosystolic murmur grade 4/6 best heard over the right midsternal border radiating to the back. The liver border was felt over the left upper quadrant. Pulses were present all over. There was grade I digital clubbing.

Chest x-ray revealed dextrocardia, collapse of the right lower lobe, a right pleural effusion, and infiltrates in the left lung field with hyperinflation. Ultrasound of the abdomen showed a left-sided liver, right-sided spleen and stomach, a pancreas in inverted orientation with the head to the right and the tail to the left, and the aorta descending to the right of the inferior vena cava, which is situated on the left of the vertebral column.

Computed tomography scan of the paranasal sinuses revealed diffuse mucosal thickening and a mass in the right nasal cavity causing left septal deviation and thinning of the medial wall of the maxillary sinuses.

Echocardiography showed dextrocardia, visceroatrial situs inversus (I), D-loop ventricles with atrioventricular discordance and ventriculo arterial discordance, D-transposition of the great arteries with anterior aorta and posterior pulmonary valve, a moderate size subpulmonary ventricular septal defect (VSD), and valvar pulmonary stenosis with a peak systolic maximum gradient of 64 mmHg. Cardiac catheterization was performed and confirmed the above described findings (Fig. 1, A, B). Sputum culture revealed heavy growth of *Hemophilus influenzae* type b.

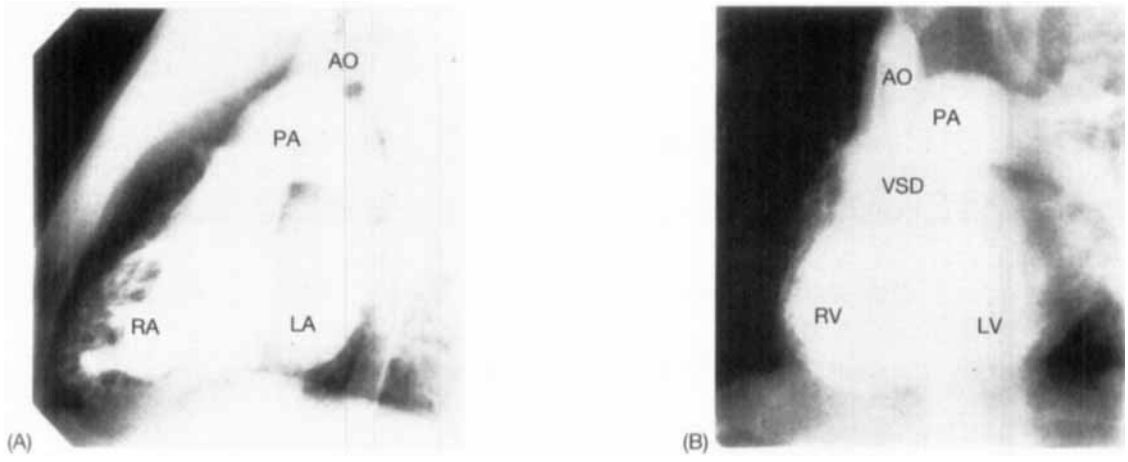

FIG. 1 Dextrocardia with physiologically "corrected" transposition of the great arteries (I,D,D). Our patient has inversus of the viscera and atria (I), with a discordant D-loop (D), and with D- TGA (D). (A) Angiogram in the lateral projection. A catheter is placed retrograde, through the aorta (Ao) into the anterior morphologic right ventricle (RV). The RV is seen emptying into the Ao and through a subpulmonary ventricular septal defect (VSD) into the pulmonary artery (PA). The PA drains into the left atrium (LA), with discordant LA-RV connection. (B) Angiogram in the anteroposterior projection. A catheter is placed retrograde across the aortic valve in the trabeculated right ventricle (RV), indicating a D-loop. Discordant RV-AO connection, with a D-transposed AO. Left ventricle (LV) and PA fill through a VSD with discordant LV-PA connection.

Pneumonia was treated and surgical repair of the cardiac lesion was successfully performed with no residual defect.

Discussion

Kartagener's syndrome is usually associated with atriovisceral situs inversus and mirror-image dextrocardia, with the segmental combination of atrial situs inversus (I), L-loop ventricles, and inverted normally related great arteries (I,L,L). No other cardiac anomalies are described more frequently than those in the normal solitus heart (S,D,S).

Transposition of the great arteries with the segmental combination of visceroatrial situs solitus (S), L-loop ventricles, and L-transposition of the great arteries (S,L,L) is the classical form of physiologically corrected transposition of the great arteries.This cardiac anomaly was the most common among 136 autopsied cases with dextrocardia from three sources (21%).[3] Associated cardiac anomalies included pulmonary outflow tract atresia, pulmonary outflow tract stenosis, VSD, single left ventricle, and others.

On the other hand, corrected transposition of the great arteries with visceroatrial situs inversus (I), D-loop ventricles, and D-transposition of the great arteries (I,D,D) is rare, occurring in only 3 of the 136 cases of dextrocardia. Although the segmental cardiac connections would theoretically be physiologically correct, the associated anomalies in the cases reported were severe, resulting in profound hemodynamic impairment.[3,4]

To our knowledge, our patient is the first reported in the literature of Kartagener's syndrome, dextrocardia, and corrected transposition of the great arteries (I,D,D), subpulmonary VSD, and valvar pulmonary stenosis.

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