Isolated Dextrocardia in a Commercial Pilot Candidate

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Positional anomalies of the heart are rare and are seldom found during routine physical examinations. We describe the case of a 25-yr-old Swiss airline pilot candidate whose aeromedical examination was normal except that an unusual ECG raised suspicion, leading to a diagnosis of dextrocardia. Further tests were conducted to rule out associated cardiac malformations, conduction anomalies, or rhythm disturbances. The patient was free of any known, pre-existing disease. Testing also excluded other associated diseases such as primary ciliary dyskinesia and Kartagener's syndrome. Dextrocardia is not listed as a disqualifying condition in the applicable aeromedical regulations (Joint Aviation Authorities, Flight Crew Licensing guidelines). Therefore, after demonstrating that there were no physical, hemodynamic, or electrophysiological abnormalities, the candidate was allowed to enroll in civilian pilot training without restrictions.

**Keywords:** aviation cardiology, commercial pilot license, European Joint Aviation Authorities, Kartagener's syndrome, dextrocardia.

**Positional Anomalies** of the heart are rarely found during routine clinical examinations (17). We encountered dextrocardia in a young man undergoing medical screening as a prerequisite for enrollment in a training program for professional civilian pilots. Additional tests were required to rule out associated anatomical or hemodynamic conditions that would be incompatible with medical certification as a pilot.

**Case Presentation**

We encountered the patient when he submitted to medical screening for JAR-FCL Medical Class 1 Licensing as a pilot. All such examinations are performed at the Institute of Aviation Medicine of the Swiss Air Force, which is the Aero Medical Center of Switzerland. He was an apparently healthy man, 25 yr of age, who reported no familial history of inherited disease and had three healthy brothers. His medical history was negative except for common childhood diseases, fractures of finger and thumb, and a right-sided herniotomy at age 20. He was physically well trained, performed well in a number of different sports, and had earned several awards in military service.

On examination, the patient was 177 cm tall, weighed 71 kg and appeared healthy. His resting BP was 120/70 mmHg and heart rate was 64 bpm. However, his electrocardiogram showed negative atrial and ventricular complexes (which are positive in Wilson III) and abnormal repolarizations in different leads, as well as an unexpected septal lateral R-wave progression (Fig. 1). The QRS-vector indicated a vertical position with right axis deviation. The precordial leads (Einthoven V1–6) were of remarkably low amplitude and showed QRS-complexes with a delayed left axis progression resembling an old septal lateral infarction. These unusual findings led to consideration of the following differential diagnosis: a) incorrect positioning of the electrodes, b) a silent myocardial infarction, or c) a cardiac/visceral positioning anomaly, in particular dextrocardia.

The correct position of the ECG electrodes was confirmed, following which a repeat ECG gave the same results. The patient was then interviewed again with a focus on his early childhood. The detailed history was negative for any known, pre-existing disease. Specifically, we ruled out cardiac or cardiovascular manifestations, obstructed nasal breathing, recurring paranasal sinusitis, and/or bronchitis. The subsequent physical examination was unremarkable except for cardiac auscultation: the point of maximal impulse was not in its usual location, but was instead shifted toward the midline and the right parasternal site. The liver edge was positioned normally and there was no sign of a splenic abnormality.

The chest X-ray (Fig. 2) confirmed the suspected atypical cardiac position (9,15). The candidate was initially surprised when the finding of dextrocardia was disclosed and the chest X-rays were shown to him, but he later recalled that he had once been told that he had a “right-sided heart.” However, in the absence of any...
Fig. 1. Standard 12-lead ECG for the patient.
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Fig. 2. The patient’s p/a chest X-ray showing isolated dextrocardia with normal position of the aorta. The normal positioning of the dia-

phragm suggests normal location of the liver. 

With normal position of the aorta. The normal positioning of the dia-

phragm suggests normal location of the liver. 

Discussion

As was the case here, asymptomatic pilot candidates may not mention a known anomaly of cardiac position in their medical history. Detection of such a condition requires careful review of all clinical data, correct positioning of ECG electrodes, and close attention to the labeling of left and right sides on images.

Cardiac orientation is determined from the base-to-
apex axis. The cardiac apex is normally located in the left side of the chest (levocardia), but may instead be found either in the right side of the chest (dextrocardia) or centrally (mesocardia). However, it is also necessary to establish the orientation of the cardiac chambers; the normal arrangement of atria and viscera is termed “situs solitus,” while its mirror image is “situs inversus.” Thus, this patient was said to have “dextrocardia with situs solitus.”

Dextrocardia with complete situs inversus occurs in approximately 2 per 10,000 live births and is accompanied by congenital heart disease (usually transposition of the great vessels) in only 3–5% of cases (18). This patient’s condition, dextrocardia with situs solitus, is less common (approximately 1 in 20,000 live births) but has more aeromedical significance because about 90% of cases have cardiovascular abnormalities, including transposition of the great arteries (50–75%), double outlet right ventricle (10–18%), ventricular septal defect (60–80%), single ventricle (15–40%), and pulmonary artery stenosis or atresia (70–80%). Polysplenia or asplenia is found in about 30% (7,6).

Because discordance between the direction of the cardiac apex and the abdominal organs suggests congenital heart disease, it is necessary to rule out the presence of congenital cardiac malformations (e.g., transposition of the great vessels, interatrial or interventricular communications) as well as conduction abnormalities and rhythm disturbances. Testing should include 2-D Doppler echocardiography (13), exercise ECG, and Holter ECG. Further testing may include cardiothoracic imaging by means of computed tomography (CT) or magnetic resonance imaging (MRI). CT provides good anatomic detail and is useful for confirming visceral organ position, cardiac apical position, and branching of the great vessels (14). MRI is particularly helpful in establishing the position of the atria (18) and is also a valuable adjunct of echocardiography in demonstrating the presence or absence of congenital heart disease. Angiography is not needed to establish the diagnosis in these cases, and is usually reserved for further evaluation where congenital cardiac malformations are found. Dextrocardia can also be associated with primary ciliary dyskinesia (PCD) and/or Kartagener’s syndrome, so the work-up of such a patient should also include biopsies of the nasal and/or bronchial mucosa along with a sperm sample (16).

PCD was first described by Afzelius et al. in 1975, and is an inherited, autosomal recessive syndrome with a prevalence of 1:15,000 to 1:35,000 without specific ethnic or gender predilection (2). The syndrome is characterized by a microtubule defect which results in immotile or ineffective ciliary movements (3); its effects can range from an isolated molecular defect without clinical significance to manifestations that include chronic respiratory problems, chronic otitis media, hydrocephalus, anosmia, and dyskinetic or akinetic spermatozoa, al-
though the latter is not necessarily linked to infertility (5).

In 1904 Siewert (19) discovered a rare congenital syndrome that was then fully described in 1933 by Kartagener (20), a specialist in Internal Medicine from Zürich, Switzerland. Kartagener’s Syndrome consists of bronchiectasis, situs inversus, and chronic sinusitis and may also include the presence of nasal polyps, an underdeveloped or absent sinus frontalis, and/or an IgA deficiency (2). Kartagener’s syndrome is a specific subgroup of PCD disorders (4,12) and is present in approximately 20% of patients with dextrocardia and situs inversus (11). It occurs with a prevalence of 1:30,000 to 1:40,000 (1).

All Swiss aviation licenses are issued in accordance with rules set forth by the European Joint Aviation Authorities. The most recent edition of the Joint Aviation Authorities medical manual, JAR FCL-3, does not specifically mention dextrocardia or situs inversus. Subpart B of section 1, paragraph 3.145 (f), does mention that applicants with any congenital abnormality of the heart shall be assessed as unfit, but may be declared fit by the Aeromedical Section following a full cardiac investigation that may include 2D Doppler echocardiography, exercise ECG and Holter ECG. The document specifies that regular cardiac review shall be required and there may be a restriction to multicrew-Class 1 OML operation.

Since the current edition of the JAR FCL-3 does not list isolated dextrocardia as a disqualifying condition, a candidate who passes exhaustive cardiac assessment can be admitted to a civilian pilot training program as Class 1 Medical qualified without restrictions. We believe that in the future, the JAR Medical Committee will need to re-evaluate the aeromedical implications of this type of anatomic variation. Cardiac assessment by the Aeromedical Section should specifically exclude the presence of associated congenital disorders such as congenital heart malformations, conduction or rhythm disturbances, primary ciliary dyskinesia, or Kartagener’s syndrome. We also suggest that specialized imaging, such as CT scan or MRI, may, in certain cases, further define the diagnosis and/or eliminate the need for more invasive diagnostic procedures. In cases in which there is a verifiable absence of additional pathology, and in which there are both normal hemodynamics and normal electrophysiology, further investigation is not needed. Although such heart anomalies are rare, they require a common evaluation policy in order to avoid inconsistencies and the potential for aeromedical tourism.

REFERENCES