JACC: CASE REPORTS VOL. 3, NO. 4, 2021

MINI-FOCUS ISSUE: ELECTROPHYSIOLOGY

ADVANCED

CASE REPORT: CLINICAL CASE

Supraventricular Tachycardia in Situs Inversus Totalis and Congenitally Corrected Transposition of the Great Arteries

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ABSTRACT

We share a rare case of an older adult patient with situs inversus totalis (mirror image of all viscera, including dextrocardia) and congenitally corrected transposition of the great arteries who presented with supraventricular tachycardia. We highlight the unique electrocardiographic characteristics of these rare conditions and the importance of electrocardiogram interpretation skills. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2021;3:597–602) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

An 80-year-old woman was conveyed by ambulance to the Department of Emergency Medicine, Sengkang General Hospital, Singapore for palpitations that started 30 min before presentation. She was tachycardic at a heart rate of 179 beats/min, with a blood pressure of 110/78 mm Hg. Physical examination was normal apart from a regular tachycardia.

LEARNING OBJECTIVES

- To understand the ECG changes in a patient with dextrocardia and CCTGA.
- To appreciate the importance of establishing the right diagnosis before starting treatment.

PAST MEDICAL HISTORY

She had a significant medical history of situs inversus totalis (this was not known during the early phase of her management). She also had a history of hiatus hernia, antral gastritis, hypertension, and a hysterectomy for uterine fibroids.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis in the emergency department was sinus tachycardia or supraventricular tachycardia (SVT). Given her age and medical history, ventricular tachycardia (VT) was also considered. Potential triggers to the conditions mentioned include infection, dehydration, and acute coronary syndrome.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received October 7, 2020; revised manuscript received January 6, 2021, accepted January 22, 2021.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2021.01.029

JACC: CASE REPORTS, VOL. 3, NO. 4, 2021
APRIL 2021:597–602

ABBREVIATIONS AND ACRONYMS

AV = atrioventricular

AVNRT = atrioventricular nodal re-entry tachycardia

AVRT = atrioventricular re-entrant tachycardia

CCTGA = congenitally corrected transposition of the great arteries

ECG = electrocardiogram

LV = left ventricle

RV = right ventricle

SVT = supraventricular tachycardia

VT = ventricular tachycardia

INVESTIGATIONS

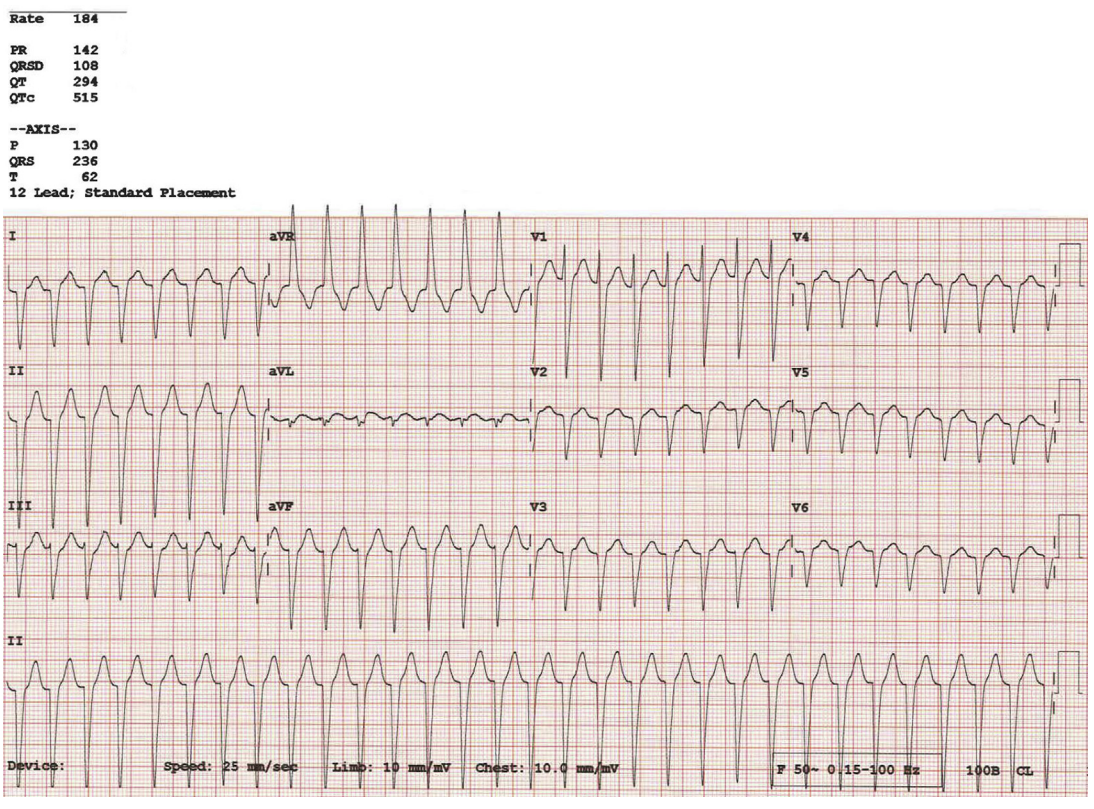
The initial electrocardiogram (ECG), which was performed with normal lead placements, showed a regular narrow complex tachycardia at 180 beats/min, a QRS complex duration of 108 ms, and no appreciable P waves (Figure 1). This result suggested that the underlying rhythm was a form of SVT favoring atrioventricular nodal re-entry tachycardia (AVNRT). However, given that a QRS complex of 108 ms approaches the upper limit of what is considered narrow complex, VT could still have been a differential diagnosis.

The QRS complex axis was very abnormal at 236° (northwest axis). It is unusual for a narrow complex tachycardia to have such an extreme axis, which is more typically observed in VT. The possibility of erroneous limb lead placement should be strongly considered in such cases.

There was abnormal R-wave progression with an rS complex in V_1 and QS complexes from V_2 to V_6, typical of patients with dextrocardia.

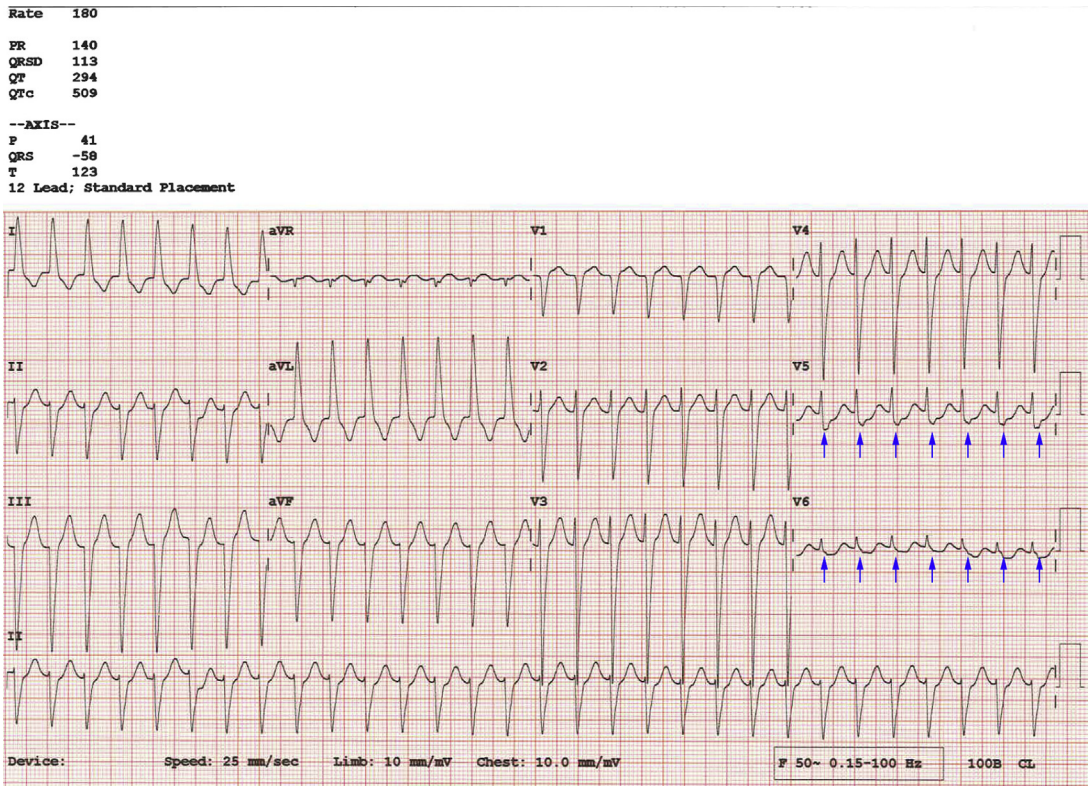
A repeat ECG with limb and precordial leads in the mirrored position was performed once the history of situs inversus totalis was known (Figure 2). This ECG showed left-axis deviation (−58°) with normal R-wave progression. The axis deviation could have been a manifestation of an anterior fascicular block. There were also retrograde P waves now seen more clearly in leads V_5 and V_6, thus indicating that this was a short RP tachycardia. This finding favored the diagnosis of AVNRT or atrioventricular re-entrant tachycardia (AVRT). AVRT is increasingly uncommon with

FIGURE 1 Initial 12-Lead Electrocardiogram



Regular narrow complex tachycardia with no obvious P waves.

FIGURE 2 Corrected 12-Lead Electrocardiogram



This electrocardiogram was obtained with leads in the mirrored position (both upper and lower limb leads were reversed) showing narrow complex tachycardia with retrograde P waves **(arrows).**

age; hence AVNRT was more likely in this 80-year-old patient.

Incidentally, there was horizontal ST-segment depression of 1 to 2 mm in leads V_5 and V_6 that could have indicated myocardial ischemia during tachycardia.

MANAGEMENT

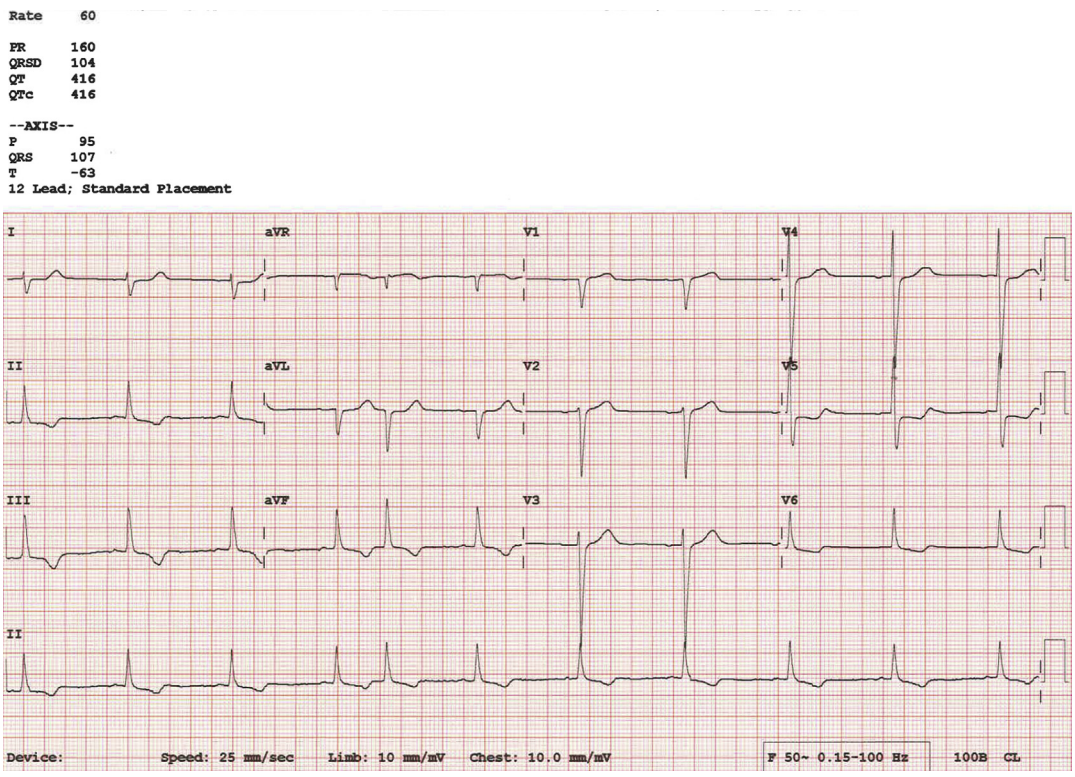
The patient was treated with 2 doses of adenosine following an unsuccessful modified Valsalva maneuver. Her SVT did not convert to sinus rhythm. She was subsequently started on an intravenous diltiazem infusion, and her cardiac rhythm reverted to sinus rhythm after 21 mg of diltiazem. The post-conversion ECG was performed with the limb and precordial leads mirrored (**Figure 3**).

The post-conversion ECG was in sinus rhythm. The positive P-wave in sinus rhythm in lead aVL on the ECG with mirrored leads indicated that atrial activation was occurring from the left to the right. This finding indicated that the sinus node was on the patient's left; hence there was atrial situs inversus. The QRS complex axis had then changed to right-axis deviation. There was an absence of R waves in lead V_1 and a loss of small Q waves in the lateral precordial leads. This finding suggested reversed septal depolarization secondary to ventricular inversion, which raised the possibility of congenitally corrected transposition of the great arteries (CCTGA) (1).

The chest radiograph demonstrated dextrocardia, with the heart in the right hemithorax and the apex pointing to the right (**Figure 4**).

The transthoracic echocardiogram obtained during her hospitalization showed that she had atrial situs inversus (inferior vena cava on the left). The morphological left atrium connected to the

FIGURE 3 Post-Conversion 12-Lead Electrocardiogram



This electrocardiogram was obtained after a total of 21 mg of diltiazem was given. The electrocardiogram leads were in the mirrored position.

morphological right ventricle (RV) (the systemic ventricle), which then connected to the aorta. The morphological right atrium connected to the morphological left ventricle (LV) (the pulmonary ventricle), which then connected to the pulmonary artery.

These findings confirmed that the patient had visceral situs inversus, dextrocardia, and CCTGA (segmental analysis: [situs inversus, dextrocardia, levo-transposition of the great arteries] discordant atrioventricular [AV] and ventriculoarterial connection).

DISCUSSION

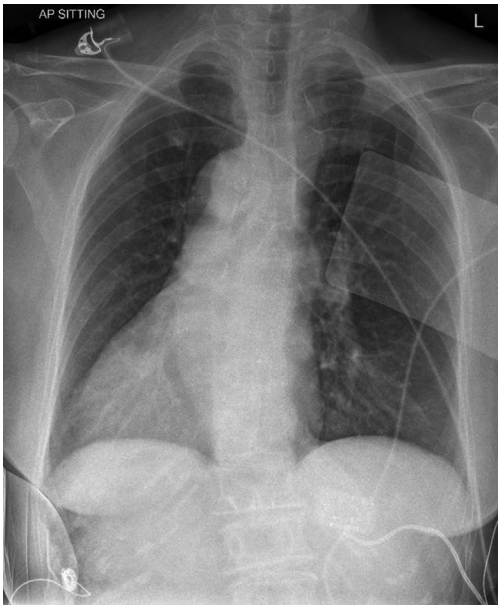
Dextrocardia is a condition in which the heart points toward the right side of the chest. The true incidence of this condition in the population is not well known, although 1 study in 2007 quoted an incidence rate of approximately 1 in 12,019 pregnancies (2). The matter is further complicated by the subcategorizations of dextrocardia, depending on whether the different chambers of the heart and the great vessels are themselves mirrored (3).

The treating physician should know the expected ECG changes when normal lead placements are used for a patient with dextrocardia. The typical findings include right-axis deviation, global inversion in lead I (downward P and T waves, with a negative QRS complex), positive QRS complex in aVR with upward P and T waves, and poor R-wave progression (4). However, one must understand that these changes may not always be present given the varying degrees of cardiac chamber malposition (5).

When dextrocardia is suspected, traditional ECG lead placements will need to be altered to obtain an accurate electrocardiographic picture of the heart (mirrored position). The limb leads will need to be reversed (left limb leads placed on the right limbs, and vice versa). Precordial leads also need to be mirrored, where V_1 now is placed over the fourth intercostal space to the left of the sternum, V_2 is at the fourth intercostal space to the right of the sternum, and V_3 to V_6 assume the positions of V_3R to V_6R.

Dextrocardia and situs inversus totalis are associated with other congenital heart defects in about 3%

FIGURE 4 Chest Radiograph



The patient's chest radiograph shows the cardiac apex on the right, the aortic knuckle on the left, and the hepatic shadow below the left hemidiaphragm. AP = anteroposterior; L = left.

to 5% of cases. The subtle absence of an R-wave in lead V_1 and the loss of small Q waves in the lateral precordial leads were clues to reversed septal depolarization, thus raising the possibility of CCTGA.

The previously undiagnosed CCTGA provided an additional dimension of complexity to this case. It is a rare condition with an estimated prevalence of 4 in 100,000 adults (6). This is a result of concurrent AV and ventriculoarterial discordance during embryologic development, leading to the morphological RV functioning as the systemic ventricle and the morphological LV functioning as the pulmonary ventricle (7).

SVT is commonly associated with CCTGA. Its association with Ebstein malformation increases the prevalence of a right-sided accessory pathway causing AVRT. The increased prevalence of AVNRT could be related to anatomic displacement of the conducting system. The presence of multiple AV nodes is also well known and can give rise to internodal AV re-entry (8).

The clinical course is variable, with a significant proportion of patients developing conduction disturbances or systemic right ventricular failure in the fourth decade of life. It is unusual for such an individual to live to 80 years of age without any disabling symptoms. The oldest recorded patient with asymptomatic CCTGA was 83 years old (9).

Generally, the expected ECG findings in SVT for these patients should resemble those in normal hearts. However, as our case demonstrates, concurrent dextrocardia in CCTGA can give rise to an abnormal cardiac axis and R-wave progression.

FOLLOW-UP

The patient was offered medication versus a more invasive approach (electrophysiological studies, coronary angiogram, and ablation). She opted for conservative management for her SVT and was discharged with an oral beta-blocker and a follow-up appointment at the adult congenital heart disease clinic.

CONCLUSIONS

When attending to an acute case in a patient with an abnormal ECG, physicians should remember the basics of ECG interpretation and carry a high index of suspicion for the potential differential diagnosis if the ECG complexes appear out of the ordinary. This vigilance will aid in providing effective and timely care for the patient.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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JACC: CASE REPORTS, VOL. 3, NO. 4, 2021
APRIL 2021:597–602

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KEY WORDS **atrioventricular nodal reentry tachycardia, congenitally corrected transposition of the great arteries, dextrocardia, ECG, supraventricular arrhythmias**