Diagnostic dilemma with a narrow QRS regular rhythm at normal rates in a patient with corrected transposition of great arteries

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A 35 year old male, known case of corrected transposition of great arteries (CTGA) and intact ventricular septum and no pulmonary stenosis, presented with history of dyspnea on exertion NYHA II and recurrent pre-syncope of 3 months duration. A baseline 12 lead electrocardiogram (ECG) at presentation (Fig. 1) showed a regular rhythm with narrow QRS at the rate of 75 beats per minute (bpm), P waves occurring on the upstroke of T waves and apparent 1:1 P-QRS relationship. The QRS shows absence of septal Q waves in I, aVL and V6 that is characteristic of CTGA.

Discussion
The ECG shows a narrow QRS regular rhythm at normal rate, but with the P wave occurring after the QRS complex. The possibilities could be considered include:
A) Complete AV block with junctional escape.
B) Junctional rhythm with 1:1 retrograde conduction.
C) Junctional rhythm with isorhythmic AV dissociation with sinus P waves
D) First degree AV block with 1:1 AV conduction.

Complete AV block with a junctional escape is the first possibility considering that the patient has underlying corrected transposition of great arteries. Complete AV block in corrected transposition of great arteries is more common with intact septum and is seen in about 52% of patients and occurs uniformly at 2% per year [1]. However, in complete AV block, the atrial rates are usually higher than the ventricular rate, and atrio-ventricular dissociation is characteristic. In this patient, the atrial rate is equal to ventricular rate with apparent 1:1 P-QRS relationship which makes complete AV block with junctional escape unlikely. Junctional rhythm with 1:1 VA conduction is also unlikely as the P waves in leads II, III and aVF are upright and not inverted [2]. The third possibility...
of junctional rhythm with isorhythmic AV dissociation needs to be considered more closely. The term AV dissociation is applied when the atrial and ventricular rhythms are independent of each other and the broad definition includes AV block which is a disorder of impulse conduction and also isorhythmic AV dissociation which is a disorder of impulse formation. Isorhythmic AV dissociation can occur by default when the primary pacemaker which is the sinus node slows down, or by usurpation when the junctional or the subsidiary pacemaker accelerates. With isorhythmic AV dissociation the rates of the dissociated pacemakers are nearly identical and the two rhythms appear to chase each other, which prompted Marriot and Menendez to describe the relationship as “flirtatious” [3]. When the relationship is persistent for a period of time it is called as “synchronization” and the RP relationship would be fixed for that duration. If the relationship is transient

![Fig. 1](image1.png)

**Fig. 1** – 12 lead ECG showing a regular rhythm with narrow QRS at the rate of 75 bpm, atrial rate of 75 bpm and P waves occurring on the upstroke of T wave with 1:1 P-QRS relationship.

![Fig. 2](image2.png)

**Fig. 2** – 24 h holter strip – when the sinus rate slows to 60 bpm, P wave is conducted with a very prolonged PR interval with 1:1 AV relationship. Note that at slower heart rates, the P waves (*) are distinctly separated from the QRS complexes.
it is called as “accrochage”, and the RP relationship would vary. The P wave morphology is normal in isorhythmic dissociation and there is changing P-QRS relationship.

One of the ways to detect changes in the P-QRS relationship is to have longer ECG strips or holter strips. If in the longer strips atrial rates are seen to be more than the ventricular rates a diagnosis of complete AV block can be made. Fig. 2 shows the holter strip of the patient when the sinus rate slows down to 60 beats per minute showing the relationship of the P to QRS complex. The P is seen to conduct 1:1 to the ventricle with a prolonged PR interval suggesting that this rhythm is not isorhythmic AV dissociation but suggests the possibility of marked first degree AV block. Note that at slower heart rates, the P waves (*) are distinctly separated from the preceding QRS complexes. The PR interval is constant at 640 ms with no evidence of AV dissociation further indicating that there is no complete AV block.

To further elucidate the AV conduction pattern, the patient underwent a diagnostic electrophysiology study prior to consideration of permanent pacemaker. Fig. 3 is an intracardiac recording obtained using three quadripolar diagnostic catheters (C.R Bard, Inc. MA, USA) positioned in the His

Fig. 3 – Intracardiac recording–shows high to low activation of the P waves (A in the HRA followed by A in His) suggesting sinus rhythm, with a prolonged AH interval of 654 ms. Note the constant 1:1 A and V relationship.

Fig. 4 – Intracardiac recording after isoprenaline showing a shorter sinus cycle length of 620 ms and a wenkeback block at the AV node.
bundle, right atrial appendage and right ventricle. It shows high to low atrial activation, prolonged AH interval of 654 ms and a normal HV interval of 36 ms. The A and V relationship is constant confirming the diagnosis of first degree AV block. On isoprenaline, sinus cycle length decreased from 840 ms to 620 ms with shortening of AH interval and development of Type 1 second degree AV block confirming AV nodal location of the block (Fig. 4).

The diagnosis of first degree AV block can easily be overlooked and misinterpreted as junctional rhythm when the PR interval is very long, with the P wave merging with the preceding QRS complex or inscribed on the preceding T wave. The diagnosis may be confirmed by a sufficiently long ECG or holter recordings or, in questionable cases, by intracardiac recordings with characteristic pattern described above. In CTGA, the position of the His is anterior and just beneath the pulmonary valve and it has been suggested that it may be difficult to record His in this anomaly [1]. Though, it has been suggested that PR intervals as long as 1000 ms may occur in first degree AV block, to our knowledge this probably is the longest documented PR interval in first degree AV block reported in literature [4]. Prolonged PR interval of more than 300 ms (0.3 s) may cause inappropriate timing of atrial and ventricular contractions resulting in hemodynamic derangement and causing symptoms mimicking a pacemaker syndrome. Such patients benefit from restoration of AV synchrony by a dual chamber pacemaker implantation [5]. The patient underwent a successful dual chamber pacemaker implantation with resolution of symptoms.

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**Conflict of interest**

None.

**References**

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