

OVERVIEW OF THE ECG

The ECG shows a wide complex tachycardia with a ventricular rate of 150bpm. Most of the QRS complexes are wide (except complexes 2 and 19 which are narrow). The QRS complexes are not regular, but occur in a regularly, irregular pattern (e.g. the cycle length between complexes 3 and 4 is shorter than between complexes 4 and 5). This repetitive pattern of short-long sequences is seen between complexes 3 and 18.

The QRS morphologies in VI are variable. Complexes 14 and 18 are identical and complexes 15 and 17 are identical. These complexes have a variable Qr and QR pattern which do not resemble a typical right bundle branch block (RBBB) morphology. Complex 16 has a QS pattern which does not resemble a typical left bundle branch (LBBB) morphology. These wide complexes cannot be conducted via the normal His-Purkinje system and must be ventricular in origin. Complex 19 is a narrow beat (100ms) with a brisk initial R wave preceded by a P wave which suggests that this beat is conducted via the normal His-Purkinje system without bundle branch block. Sinus tachycardia with RBBB with ventricular bigeminy, and alternating left anterior and posterior fascicular block, can therefore be excluded. QRS morphologies are not expected to change to this degree with variable degrees of pre-excitation and the regularly, irregular pattern excludes pre-excited atrial fibrillation.

In the limb leads, the complexes can be seen to alternate in morphology and axis. QRS complexes 3, 5, 7, 9, 11, 13, 15, 17, 21, 23 and 24 appear identical and are superiorly directed (-75 degrees) and QRS complexes 4, 6, 8, 10, 12, 14, 16, 18, 22 and 25 appear identical and are inferiorly directed (+90 degrees). The tachycardia stops with complex 18 and starts again with complex 20. P waves are not visible before the QRS complexes (except for complex 19). Dissociated P waves are visible before complexes 9 and 15. The presence of AV dissociation in the setting of a wide complex tachycardia confirms the diagnosis of ventricular tachycardia (VT). Slight variation in the QRS morphologies probably reflect subtle degrees of fusion between the VT and underlying conducted beats (e.g. complexes 2 and 6).

Ventricular tachycardia can be classified into 2 distinct forms: (1) Monomorphic VT or (2) Polymorphic VT (Torsade de Pointes in the setting of a long QT interval). Monomorphic VT, as its name implies, has a single stable QRS morphology from

beat to beat and is usually due to a scar re-entrant mechanism which results in ventricular depolarisation arising from a single site. Polymorphic VT has a continuously changing, or polymorphic, morphology with no constant morphology, no clear isoelectric baseline between complexes and no single site of origin. This ECG has distinct morphology QRS complexes and therefore polymorphic VT is excluded.

The correct answer is (4) Bidirectional Ventricular Tachycardia.

BIDIRECTIONAL VENTRICULAR TACHYCARDIA

This patient has an unusual VT called bidirectional VT which is associated with beat to beat alternans in the QRS frontal plane axis. The most characteristic ECG pattern is an atypical RBBB superior axis alternating with an atypical RBBB inferior axis morphology. Occasionally atypical LBBB alternating with atypical RBBB beats can be seen. The mechanism of the bidirectional VT appears to be due to a "ping-pong" mechanism of reciprocating bigeminy where a delayed after depolarisation-triggered beat from the left anterior fascicle (inferiorly directed beat) induces a triggered beat from the left posterior fascicle (superiorly directed beat) and so on (Figure 1).⁽¹⁾ Reciprocating bigeminy from 3 or more sites produce polymorphic VT.

Bidirectional VT was first described in 1922 as a manifestation of digoxin toxicity. It has also been described in other conditions like hypokalaemic periodic paralysis, Andersen-Tawil syndrome (Long QT syndrome [type 7] which is associated with periodic paralysis and facial and limb dysmorphism), fulminant and subacute myocarditis. Another example of bidirectional VT secondary to subacute myocarditis is shown in Figure 2.⁽²⁾ In the setting of a normal general and cardiovascular examination, bidirectional VT is most likely due to catecholaminergic polymorphic ventricular tachycardia (CPVT) and the resting ECG will be normal.

The correct answer is (3) "Normal" ECG.

CATECHOLAMINERGIC POLYMORPHIC VENTRICULAR TACHYCARDIA

CPVT is a genetic disease caused by mutations in the cardiac ryanodine receptor (RyR2) or calsequestrin (CASQ2) gene.⁽³⁾ The prevalence of the disease is estimated to be 1:10 000 in the general population. The typical clinical presentation consists

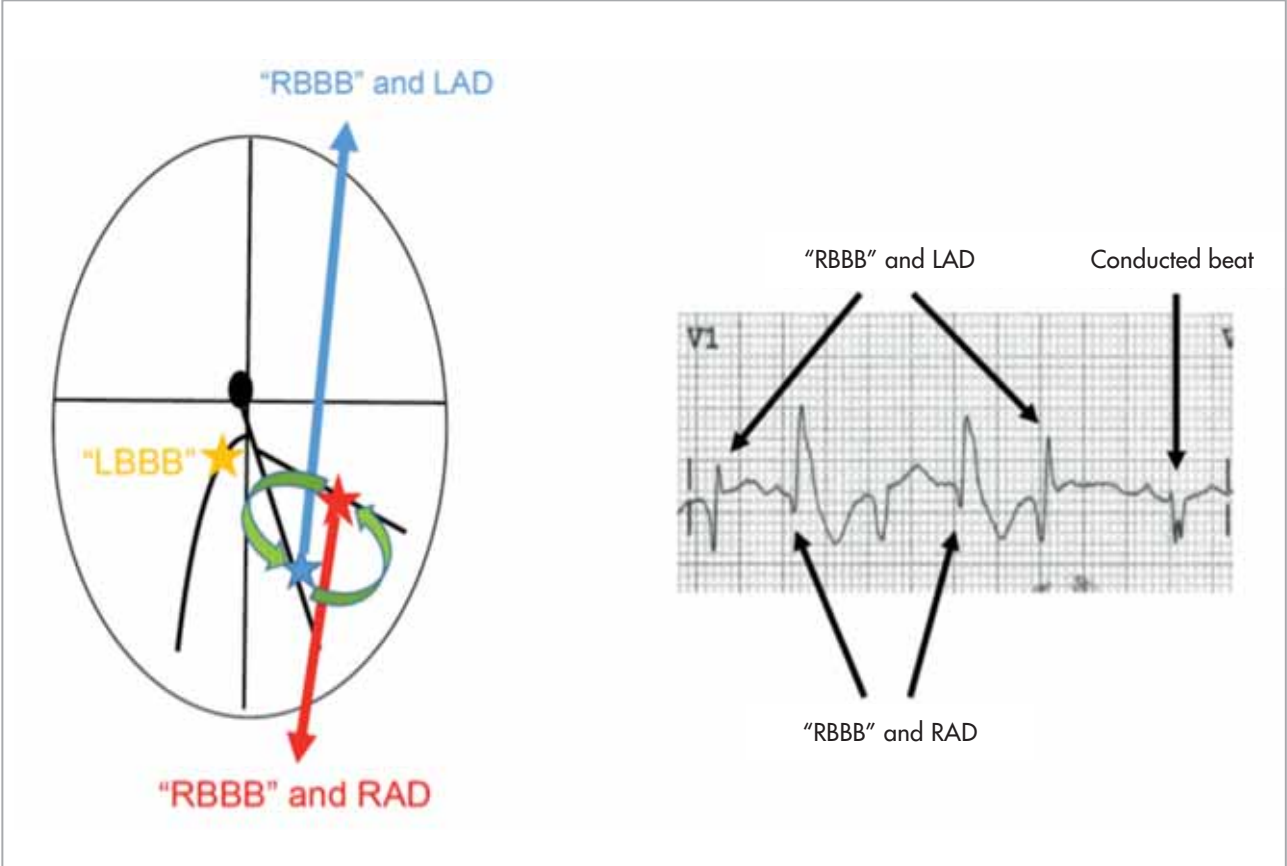


FIGURE 1: Bidirectional VT caused by reciprocating bigeminy in the left anterior and posterior fascicles.
 Ectopy arising from the left posterior fascicle (blue) produces a PVC with an atypical RBBB pattern with left axis deviation. Ectopy arising from the proximal left bundle branch or right bundle branch (yellow) produces a PVC with an atypical LBBB pattern.
 "RBBB": atypical right bundle branch block, "LBBB": atypical left bundle branch block, LAD: left axis deviation, RAD: right axis deviation

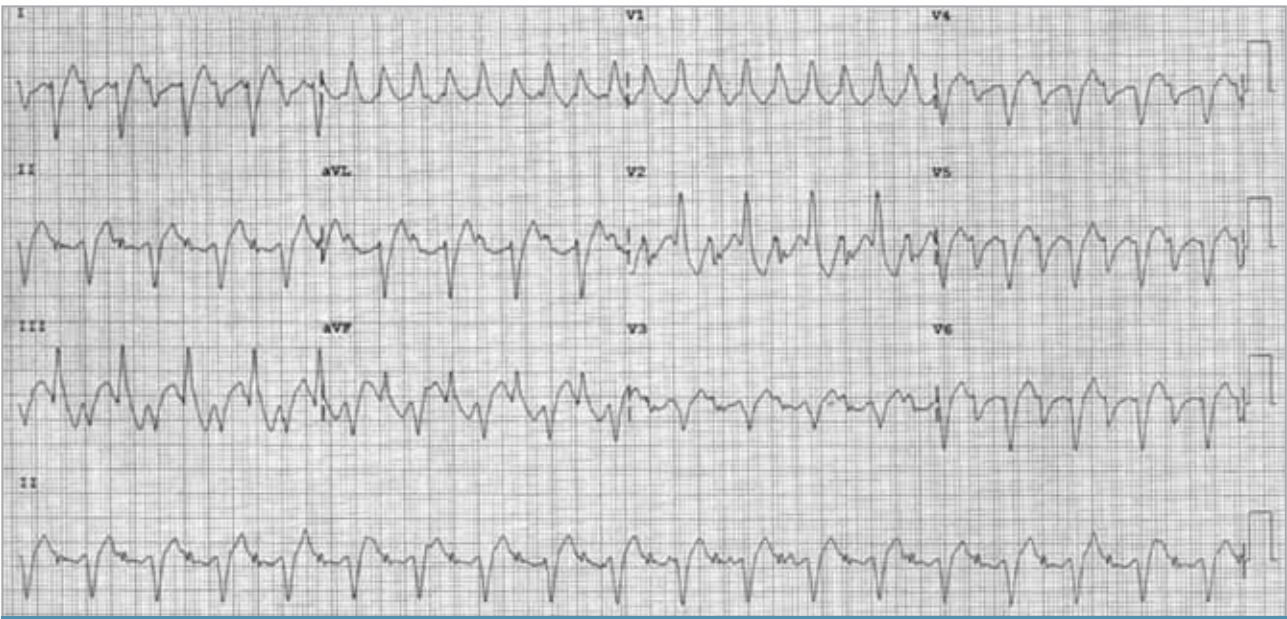


FIGURE 2: Bidirectional VT due to subacute myocarditis.
 Adapted with permission from Reference 2.



FIGURE 3: Exercise stress test diagnostic of CPVT.

Bidirectional PVCs can be seen in the limb leads. The PVCs have an atypical, variable RBBB morphology in V1 with a single beat with an atypical LBBB morphology.

of syncope, aborted cardiac arrest or sudden death during physical or emotional stress. Patients typically present in childhood, adolescence or in their 20s. Patients usually have normal resting ECGs including a normal QT interval. The diagnosis of CPVT depends on provocative testing with an exercise stress test or an adrenaline infusion. Isolated PVCs are initially seen followed by bigeminal PVCs, polymorphic couplets, non-sustained VT and bidirectional VT. The arrhythmias rapidly abate during rest.

This patient had an exercise stress test diagnostic of CPVT (Figure 3). She developed PVCs that occurred with increasing frequency during the test. She developed ventricular bigeminy with bidirectional couplets. She then developed bidirectional PVCs at peak exercise. The bidirectional PVCs had an atypical RBBB morphology in V1 and bidirectional PVCs can be seen in the limb leads. During the resting phase the ventricular ectopy became less frequent and then disappeared.

The management of CPVT includes the use of beta-blockers and the avoidance of competitive exercise. An implantable cardioverter defibrillator is indicated in patients with an aborted cardiac arrest, or in those with ventricular arrhythmias despite beta-blocker therapy. The addition of flecainide to beta-blocker therapy has been shown to be effective as second-line therapy. Left cardiac sympathetic denervation has also been shown to be effective.

CONCLUSION

Bidirectional VT is an uncommon arrhythmia that has a limited differential diagnosis. Causes include digoxin toxicity and CPVT.

The mechanism of bidirectional VT is reciprocating bigeminy in the His-Purkinje system.

Bidirectional VT usually has an atypical RBBB morphology in V1 with alternating beats showing marked changes in QRS frontal axis.

CPVT should be considered in any patient with exercise-induced syncope or cardiac arrest with a structurally normal heart and normal baseline ECG.

REFERENCES

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