

## Country cardiograms case 41: Answer

*Brent M. McGrath,  
MD, MSc, PhD  
Department of Medicine,  
Faculty of Medicine, Dalhousie  
University, Halifax, NS*

*Jeff Moore, MD,  
FRCPC  
Department of Medicine,  
Dr. Everett Chalmers  
Regional Hospital,  
Fredericton, NB*

**I**nterpretation of the electrocardiogram (ECG) shown in Figure 1 (on page 133) reveals a sinus arrhythmia — most likely due to changes with respirations — at a rate of about 70 beats/min. Of note is the shortened PR interval (80 [normal 120–200] ms). The QRS is normal in appearance and duration (81 [normal 80–120] ms). The corrected QT interval is also normal (395 ms). No other abnormalities are noted on the ECG.

The clinical presentation and ECG are diagnostic of Lown–Ganong–Levine (LGL) syndrome.<sup>1</sup> Diagnosis of LGL syndrome requires a triad of paroxysmal tachycardia, shortened PR interval and normal QRS duration. It is one of several pre-excitation syndromes, the most common being Wolff–Parkinson–White (WPW) syndrome.<sup>2,3</sup> An isolated short PR interval is relatively common, with a reported prevalence of 2%–4%.<sup>4</sup> In their 1952 study, Lown and colleagues reported tachycardia in 10% of the patients with a shortened PR interval and normal QRS duration.<sup>1</sup> Thus, the frequency of LGL syndrome can be estimated at between 0.2% and 0.4%. It typically occurs in women who are otherwise healthy (71% of cases), who range in age from 10 to 61 years.<sup>1</sup>

The ECG findings in LGL syndrome purportedly result from either an accessory pathway (i.e., para- or extranodal) between atrium and ventricles or an enhanced atrioventricular nodal conduction system (i.e., intranodal). There are several proposed accessory pathways in the syndrome; however, no single anatomic anomaly has been consistently reported. It appears more likely that several distinct

structural conduction abnormalities can all lead to this ECG presentation.

Irrespective of the precise location of the accessory pathway, this syndrome differs physiologically from WPW syndrome in the following manner. In LGL syndrome, conduction through the accessory pathway must bypass the intrinsic delay of the atrioventricular node (to produce the shortened PR interval) but it still must activate the ventricles via the bundle of His (to produce a normal QRS complex). In WPW syndrome, the PR interval is shortened by the same mechanism; however, the QRS complex is broad with the classic delta wave. This difference results from the location of the accessory pathway (i.e., the Kent bundle) in WPW syndrome. The Kent bundle bypasses both the atrioventricular node and bundle of His, thus depolarizing the ventricles directly and slightly earlier than the physiologic pathway, producing the delta wave on the ECG. Although they are physiologically different, both syndromes predispose the patient to re-entrant dysrhythmias, including atrioventricular re-entrant tachycardia and atrial fibrillation.

The evidence base is limited with respect to the management of LGL syndrome and associated tachycardia. In patients without atrial fibrillation, some studies have suggested efficacy with  $\beta$ -blockers or calcium channel blockers.<sup>5,6</sup> Cardiac glycosides, including digitalis, have not been shown to be effective.<sup>7</sup> There is theoretical rationale for class I or III antiarrhythmic agents — particularly in the context of atrial fibrillation in a stable patient — but no studies are available. As with WPW

syndrome, cardioversion is the preferred treatment modality in the unstable tachycardiac patient with LGL syndrome. As in WPW syndrome, radiofrequency catheter ablation plays a central role in treatment for patients who are experiencing frequent bothersome or dangerous tachycardia that is unresponsive to pharmacotherapy or for patients who wish to avoid long-term treatment.<sup>8</sup>

Our patient is presently experiencing several episodes of palpitations per month; however, none have been confirmed on ECG. Moreover, none have persisted long enough to enable the patient to present to a local medical clinic or hospital for an ECG. She underwent an exercise stress test to determine whether an event could be precipitated. The patient completed 15 minutes (17.2 MET) of a Bruce protocol and reached target heart rate; however, no dysrhythmia was noted and the patient's symptoms were not reproduced. No further investigation is planned at this time, because the patient's symptoms have become less frequent. However, a reasonable next step would be repeat monitoring with a Holter monitor, a King of Hearts (cardiac event) monitor (Instromedix) or an electrophysiology study. Confirmation and characterization of a tachydysrhythmia is essential before consideration can be given to pharmacologic or ablative treatment. The only

report of sudden cardiac death in LGL syndrome was in 2 patients with paroxysmal atrial fibrillation in the original study by Lown and colleagues.<sup>1</sup>

**For the question, see page 133.**

#### REFERENCES

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## Cardiogrammes ruraux

Avez-vous eu à décrypter un ECG particulièrement difficile récemment?

Dans la plupart des numéros du *JCMR*, nous présentons un ECG assorti de questions.

Les réponses et une discussion du cas sont affichées sur une autre page.

Veillez présenter les cas, accompagnés d'une copy de l'ECG, à Suzanne Kingsmill, rédactrice administrative, *JCMR*, 45, boul. Overlea, C. P. 22015, Toronto (Ontario)

M4H 1N9 ; [cjrm@cjrm.net](mailto:cjrm@cjrm.net)