



## Cardiac conduction defects and complete heart block in myotonic dystrophy

### Defectos de conducción cardiaca y bloqueo cardiaco completo en la distrofia miotónica

Sandeep Khanna<sup>a,b,c</sup>, Sergio Bustamante<sup>a</sup>

<sup>a</sup> Department of Cardiothoracic Anesthesiology, Anesthesiology Institute, Cleveland Clinic Foundation, Cleveland, Ohio

<sup>b</sup> Department of General Anesthesiology, Anesthesiology Institute, Cleveland Clinic Foundation, Cleveland, Ohio

<sup>c</sup> Department of Outcomes Research, Anesthesiology Institute, Cleveland Clinic Foundation, Cleveland, Ohio.

Classic myotonic dystrophy is a multisystem disorder that results from RNA toxicity and is one of the commonest adult onset muscular dystrophies. Patients often present with muscle stiffness from myotonia and dysphagia or dysarthria from laryngopharyngoesophageal muscle

weakness. Benign electrocardiogram changes such as first degree atrioventricular block are commonly present and rarely merit further work up. Occasionally, patients develop advanced conduction defects which can unexpectedly progress to complete heart block perioperatively.<sup>1</sup> This

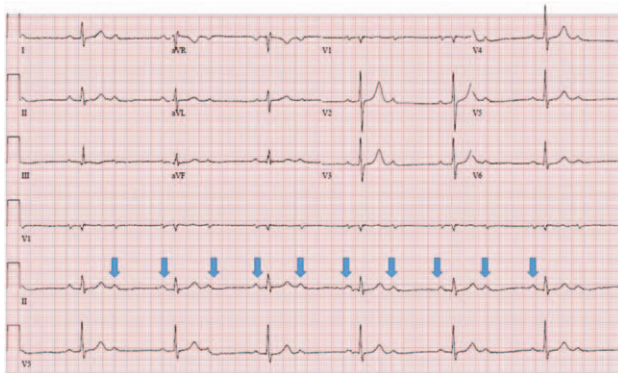


Figure 1. 2:1 Atrioventricular block.

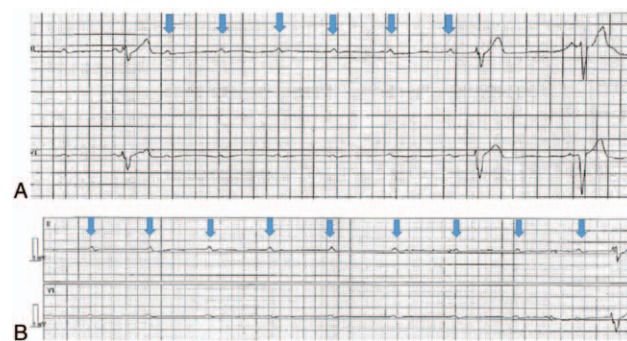


Figure 2. Complete heart block.

How to cite this article: Khanna S, Bustamante S. Cardiac conduction defects and complete heart block in myotonic dystrophy. Colombian Journal of Anesthesiology. 2020;48:162-163.

Read the Spanish version of this article at: <http://links.lww.com/RCA/A923>

Copyright © 2020 Sociedad Colombiana de Anestesiología y Reanimación (S.C.A.R.E.). Published by Wolters Kluwer. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

Correspondence: Department of Cardiothoracic Anesthesiology; Department of General Anesthesiology; and Department of Outcomes Research, Anesthesiology Institute, Cleveland Clinic Foundation, 9500 Euclid Avenue, E3-108, Cleveland, OH 44122.

E-mail: [khannas@ccf.org](mailto:khannas@ccf.org)

Colombian Journal of Anesthesiology (2020) 48:3

<http://dx.doi.org/10.1097/CJ9.0000000000000155>

circumstance is demonstrated by the accompanying images. The preprocedure 12 lead electrocardiogram exhibits a 2:1 atrioventricular block pattern (Fig. 1). The blue arrows in Lead II mark the position of P waves. For every 2 P waves, 1 narrow QRS complex is seen. During cystoscopy under general anesthesia, the rhythm suddenly deteriorates to complete heart block (Fig. 2A and B).

2:1 atrioventricular block is a distinct second-degree heart block that merits immediate consultation as even asymptomatic patients may need permanent pacemaker implantation before surgery. Presence of wide QRS complexes with this pattern suggests infra-atrioventricular nodal disease and a heightened risk of progression to complete heart block. Rarely, even narrow QRS complexes may represent infranodal disease, as in this instance. Unlike a resting electrocardiogram, a cardiac electrophysiological study reliably identifies infranodal disease.<sup>2</sup>

Hyperkalemia, hypothermia, and use of vagomimetic medications like remifentanyl or beta blockers can worsen atrioventricular conduction and precipitate complete heart block in these patients.<sup>3</sup> Hemodynamically unstable complete heart block is characterized by non-conduction of atrial impulses to the ventricles, an unreliable slow ventricular escape rhythm, abrupt decrease in cardiac output, precipitous hypotension, and cardiovascular collapse. As complete heart block is often resistant to treatment with atropine or inotropes, transcutaneous pacing should be instituted early. Persistent complete heart block necessitates transvenous pacing. Intraoperatively, invasive arterial monitoring, plethysmography, and echocardiography help confirm successful mechanical capture during pacing.<sup>2</sup>

### Ethical responsibilities

**Protection of people and animals:** no experiments on people or animals were done.

**Confidentiality of the data:** all protocols at our institute were followed and patient or hospital identifiers have been removed from all images.

**Right to privacy and informed consent:** as patient and hospital identifiers have been removed, no informed consent was solicited for this production.

### Acknowledgements

None.

### Author's contributions

Author 1 (SK): conception of project, planning, and final writing of manuscript.

Author 2 (SB): conception of project, approval of manuscript.

### Financial support

The authors have no funding to disclose.

### Conflicts of interest

The authors have no conflicts of interest to disclose.

### References

1. Russell SH, Hirsch NP. Anaesthesia and myotonia. *Br J Anaesth* 1994;72:210-216.
2. Kusumoto FM, Schoenfeld MH, Barrett C, et al. ACC/AHA/HRS guideline on the evaluation and management of patients with bradycardia and cardiac conduction delay: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. *Circulation* 2019;140:e382-e482.
3. Zaballos M, Jimeno C, Almendral J, et al. Cardiac electrophysiological effects of remifentanyl: study in a closed-chest porcine model. *Br J Anaesth* 2009;103:191-198.