

## IMAGE IN CARDIOLOGY

# Anomalous insertion of the papillary muscle in a patient with sickle cell disease: A normal variant with no left ventricular outflow obstruction

## Inserção anómala do músculo papilar num doente com drepanocitose: Uma variante do normal sem obstrução do tracto de saída do ventriculo esquerdo

Inês Zimbarra Cabrita<sup>a,\*</sup>, Julia Grapsa<sup>a</sup>, David Dawson<sup>a</sup>, Fausto J. Pinto<sup>b,c</sup>,  
J. Simon R. Gibbs<sup>a</sup>, Petros Nihoyannopoulos<sup>b</sup>

<sup>a</sup> National Heart and Lung Institute, Hammersmith Campus, Imperial College London, United Kingdom

<sup>b</sup> Instituto Cardiovascular, Lisboa, Portugal

<sup>c</sup> Hospital de Santa Maria, Centro Académico de Medicina de Lisboa, Serviço de Cardiologia I, CCUL, Lisboa, Portugal

Received 19 March 2012; accepted 28 March 2012

Available online 16 September 2012

### Introduction

Abnormalities of the papillary muscle (PM) are a rare disorder that has been described in patients with hypertrophic cardiomyopathy<sup>1–3</sup> or cardiac amyloidosis causing significant left ventricular outflow tract (LVOT) obstruction. These anomalies are often challenging to diagnose with 2D echocardiography, and real-time 3D echocardiography (RT3D) facilitates the diagnosis by providing accurate imaging and delineation of the spatial relationship between the cardiac structures.

### Case report

A 43-year-old black African woman, asymptomatic at presentation (blood pressure 105/71 mmHg, heart rate 82 bpm, body surface area 1.52 m<sup>2</sup>), diagnosed with sickle cell disease, was investigated for pulmonary hypertension in our department. Echocardiography was requested for the

assessment of cardiac function and estimation of pulmonary artery pressures.

Full 2D and 3D transthoracic echocardiography (TTE) was performed and 3D images were acquired and analyzed using Philips Qlab offline analysis software. The echocardiogram revealed a dilated left ventricle (LV) with normal wall thickness and good global systolic function.

In parasternal long-axis 2D view, the anterolateral PM appeared in an unusual position, and in short-axis view and apical 4 chambers view (Figure 1) it was clearly inserted lower than normal. A complete Doppler assessment revealed no high velocity or turbulent flow pattern and no significant LVOT or intracavitary gradient was identified.

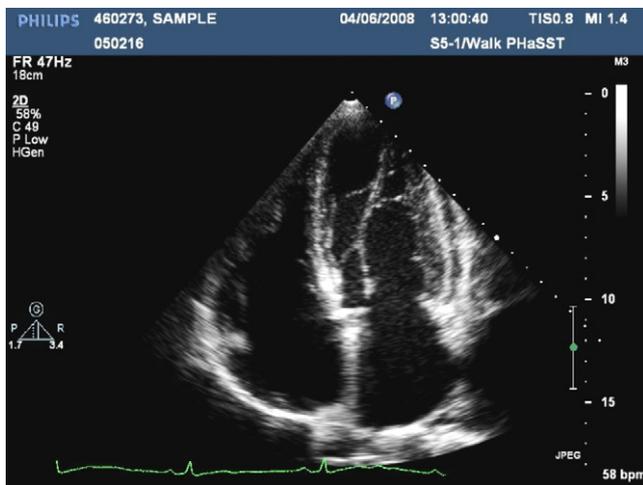
3D echocardiography clearly showed the lower insertion of the PM and the hypertrophied chordae (Figure 2). A follow-up echocardiography study was recommended to assess left ventricular hypertrophy but the patient died a few months later due to a subarachnoid hemorrhage. No post-mortem was performed.

### Discussion

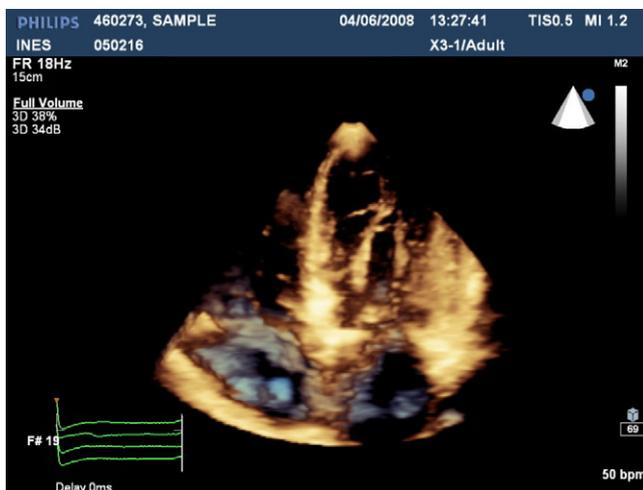
Anomalous insertion of PMs not causing LVOT obstruction is a rare abnormality.<sup>1–4</sup> RT3D is being increasingly used in the

\* Corresponding author.

E-mail address: ines.cabrira05@imperial.ac.uk (I. Zimbarra Cabrita).



**Figure 1** Apical 4-chamber view of the anomalous insertion of the papillary muscles in 2D transthoracic echocardiography.



**Figure 2** Real-time 3D echocardiography demonstrating the low insertion of the anterolateral papillary muscle in apical 4-chamber view.

clinical setting to accurately identify cardiac structures and anatomy.<sup>5,6</sup> Its role is mainly in providing additional spatial information to 2D TTE in the identification of complex anatomic structures. The normal variant of low insertion of the anterolateral PM without LVOT obstruction may be observed in healthy individuals but an association with sickle cell disease cannot be excluded; further investigations are needed to confirm this association. To our knowledge, this is the first case in sickle cell disease of an

anomalous insertion of PMs not causing LVOT obstruction reported in the literature.

## Ethical disclosures

**Confidentiality of data.** The authors declare that they have followed the protocols of their work center on the publication of patient data and that all the patients included in the study have received sufficient information and have given their informed consent in writing to participate in the study.

**Right to privacy and informed consent.** The authors have obtained the informed consent of the patients and/or subjects mentioned in the article. The author for correspondence is in possession of this document.

**Protection of human and animal subjects.** The authors declare that no experiments were performed on humans or animals for this investigation.

## Conflicts of interest

The authors have no conflicts of interest to declare.

## Appendix A. Supplementary material

Supplementary data associated with this article can be found, in the online version, at [doi:10.1016/j.repc.2012.03.004](https://doi.org/10.1016/j.repc.2012.03.004).

## References

1. Austin BA, Kwon DH, Smedira NG, et al. Abnormally thickened papillary muscle resulting in dynamic left ventricular outflow tract obstruction: an unusual presentation of hypertrophic cardiomyopathy. *J Am Soc Echocardiogr.* 2009;22:e5–6, 105.
2. Nomura T, Harada Y, Suzaki Y, et al. Left ventricular outflow tract obstruction due to anomalous insertion of papillary muscle. *Circ J.* 2004;68:1219–22.
3. Klues HG, Maron BJ, Dollar AL, et al. Diversity of structural mitral valve alterations in hypertrophic cardiomyopathy. *Circulation.* 1992;85:1651–60.
4. Watanabe N, Wada N, Yoshida K. Uncommon anomalous papillary muscle/chordae tendineae incidentally found in patient with transient ischaemic attack. *Heart.* 2005;91:p1074.
5. Yang HS, Lee KS, Chaliki HP, et al. Anomalous insertion of the papillary muscle causing left ventricular outflow obstruction: visualization by real-time three-dimensional echocardiography. *Eur J Echocardiogr.* 2008;9:855–60.
6. Hung J, Lang R, Flachskampf F, et al. 3D echocardiography: a review of the current status and future directions. *J Am Soc Echocardiogr.* 2007;20:213–33.