

UC Irvine

Clinical Practice and Cases in Emergency Medicine

Title

Image to Fit the Clinical Picture: Point-of-care Ultrasound Assessment of Ebstein's Anomaly in Peru

Permalink

<https://escholarship.org/uc/item/8qm9w8jg>

Journal

Clinical Practice and Cases in Emergency Medicine, 4(2)

Authors

Rider, Ashley C.
Dreyfuss, Andrea
Inga, Roberto

Publication Date

2020

DOI

10.5811/cpcem.2019.12.44783

Supplemental Material

<https://escholarship.org/uc/item/8qm9w8jg#supplemental>

Copyright Information

Copyright 2020 by the author(s). This work is made available under the terms of a Creative Commons Attribution License, available at <https://creativecommons.org/licenses/by/4.0/>

Peer reviewed

Image to Fit the Clinical Picture: Point-of-care Ultrasound Assessment of Ebstein's Anomaly in Peru

Ashley C. Rider, MD*
Andrea Dreyfuss, MD, MPH*
Roberto Inga, MD†

*Highland Hospital, Alameda Health System, Oakland, California
†Hospital Nacional Dos de Mayo, Department of Emergency Medicine, Lima, Peru

Section Editor: Rick A. McPheeters, DO

Submission history: Submitted August 1, 2019; Revision received November 30, 2019; Accepted December 18, 2019

Electronically published February 24, 2020

Full text available through open access at http://escholarship.org/uc/uciem_cpem

DOI: 10.5811/cpem.2019.12.44783

Ebstein's anomaly is a congenital heart defect that when left untreated can lead to unique physical exam and ultrasound findings. This case describes a patient who presented with dyspnea and was found to have cyanosis, clubbing, and dilation of right-sided chambers on point-of-care ultrasound. The series of images highlights findings in late-stage Ebstein's anomaly and serves as a springboard for the discussion of the pathophysiology, diagnosis, and treatment of this rare congenital heart disease. [Clin Pract Cases Emerg Med. 2020;4(2):222–224.]

CASE PRESENTATION

A 20-year-old male presented to the emergency department with progressive dyspnea. He was noted to have hypoxemia, clubbing of the fingers (Image 1), and perioral cyanosis (Image 2). Point-of-care ultrasound revealed a severe anatomic abnormality of the heart consistent with Ebstein's anomaly (Image 3 and video).

DISCUSSION

Ebstein's anomaly is caused by a congenital insufficiency of the tricuspid valve due to the apical displacement of the annulus. This leads to a dilated atrium and atrialization of the right ventricle as seen in this ultrasound image of a standard apical 4-chamber view.¹ Other cardiac anomalies are commonly associated, such as atrial septal defect and ventricular septal defect.²

Ebstein's anomaly accounts for less than 1% of congestive heart failure (CHF) and varies in severity.³ If tricuspid regurgitation is severe, symptoms such as CHF and cardiomegaly may develop in the neonatal period.¹ Mild cases of Ebstein's anomaly may remain undiagnosed until late childhood or adulthood, when presenting symptoms may include cyanosis and decreased exercise tolerance, as with this case. Adults also have a high risk of atrial tachyarrhythmia and ventricular pre-excitation, which predisposes patients to lethal arrhythmias.⁴

Patients with Ebstein's anomaly may require medical or surgical treatment for atrialization of the right ventricle.⁵ Medical

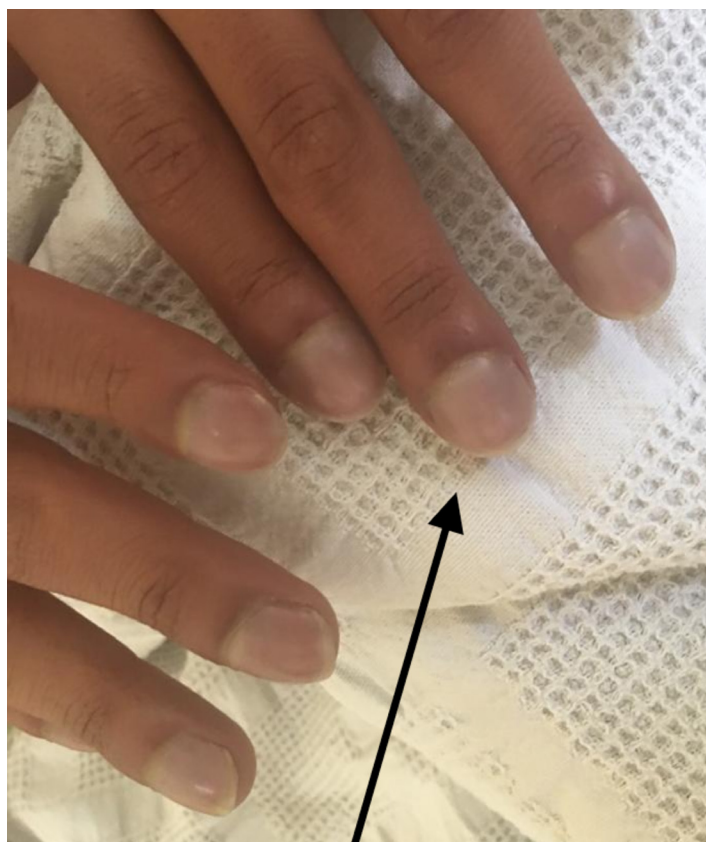


Image 1. Fingers of patient (arrow) demonstrating cyanosis and clubbing.



Image 2. Mild cyanosis of lips (arrow) demonstrating chronic hypoxemia.

treatment includes diuresis, angiotensin-converting enzyme inhibitors, and digoxin. Tricuspid valve repair or replacement may be indicated in patients experiencing deteriorating exercise capacity, cyanosis (finger oxygen saturation <90%), paradoxical embolism, cardiomegaly, or reduction of right heart function.⁵ Surgical intervention should not be delayed until right heart failure occurs as this is associated with poor outcomes.⁴ Most cases of Ebstein's anomaly fare well, especially when surgically corrected, with the majority of patients living to at least age 60.³

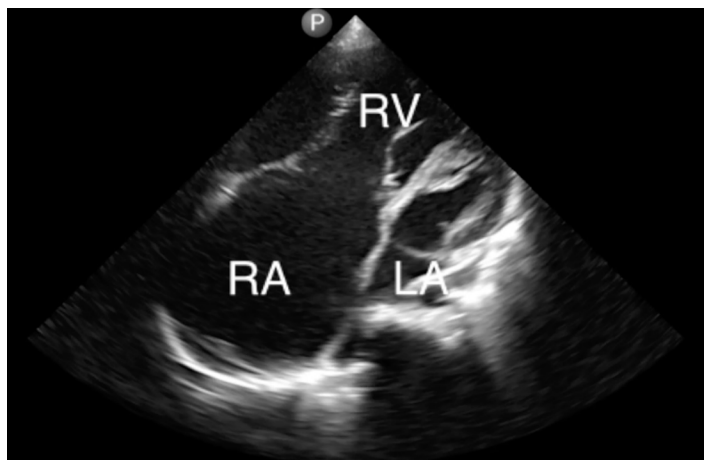


Image 3. Point-of-care ultrasound apical 4-chamber view of patient with Ebstein's anomaly demonstrating the dilated right heart chambers. The left ventricle is demonstrated by the arrow. RV, right ventricle; RA, right atrium; LA, left atrium.

Video. An apical 4-chamber cardiac ultrasound obtained in a patient with Ebstein's anomaly demonstrating the dilated right-sided chambers. The left ventricle is demonstrated by the arrow. RV, right ventricle; RA, right atrium; LA, left atrium arrow.

Documented patient informed consent and/or Institutional Review Board approval has been obtained and filed for publication of this case report.

CPC-EM Capsule

What do we already know about this clinical entity?

Ebstein's anomaly is a form of congenital heart disease caused by insufficiency of the tricuspid valve, leading to a dilated atrium and atrialization of the left ventricle.

What is the major impact of the image(s)?

These images show physical exam findings and point-of-care ultrasound (POCUS) features of late-stage Ebstein's anomaly in a patient in Peru.

How might this improve emergency medicine practice?

In settings with limited access to pediatric cardiac surgery, patients may present with late manifestations of the disease. POCUS ultrasound may help in the diagnosis.

Address for Correspondence: Ashley C. Rider, MD, Highland Hospital, Alameda Health System, Department of Emergency Medicine, 1411 E. 31st Street, Oakland, CA 94602. Email: ashley.christine.rider@gmail.com.

Conflicts of Interest: By the CPC-EM article submission agreement, all authors are required to disclose all affiliations, funding sources and financial or management relationships that could be perceived as potential sources of bias. The authors disclosed none.

Copyright: © 2020 Rider et al. This is an open access article distributed in accordance with the terms of the Creative Commons Attribution (CC BY 4.0) License. See: <http://creativecommons.org/licenses/by/4.0/>

REFERENCES

1. Attenhofer Jost CH, Connolly HM, Dearani JA, et al. Ebstein's anomaly. Congenital heart disease for the adult cardiologist. *Circulation*. 2007;115(2):277-85.
2. Luu Q, Choudhary P, Jackson D, et al. Ebstein's anomaly in those surviving to adult life: a single centre experience. *Heart Lung Circ*. 2015;24(10):996-1001.
3. Choudhary P, Luu Q, Caniffe C, et al. Abstract 18937: Ebstein's anomaly in adults: favourable outcomes from a single specialist centre experience. *Circulation*. 2018;130(suppl 2):A18937.
4. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC Guideline for

the Management of Adults with Congenital Heart Disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *Circulation*. 2019;139(14):e698-800.

5. Riaz KR and Ali YS. Ebstein anomaly guidelines. Available at: <https://emedicine.medscape.com/article/154447-guidelines>. Accessed June 17, 2019.