Too big for echocardiography

Clement Karsenty1,2,3*, Magalie Ladouceur1,2,4, Mikael Laredo1,2, Laurence Iserin1,2, and Gilles Soulat1,2

1Adult Congenital Heart Disease Unit, Department of Cardiology, Hôpital Européen Georges Pompidou, France; 2Centre de référence des malformations Cardiaques Congénitales Complexes, M3C, Paris Descartes University, Paris, France; 3INSERM UMR1048, Institut des Maladies Métaboliques et Cardiovasculaires, DMC, Equipe 8, Toulouse, France; and 4Paris Centre de Recherche Cardiovasculaire, INSERM U970, Paris, France

* Corresponding author. Tel: +33 673407752, Fax: +33 156095466, Email: clement.karsenty@hotmail.fr

A 46-year-old male was referred to our tertiary hospital for specialized follow-up. In his medical notes, there was a notion of aortopulmonary window (APW) complicated by Eisenmenger syndrome. He remained stable in New York Heart Association Class II with relatively preserved quality of life. On physical examination, digital clubbing and peripheral cyanosis were noted. The resting capillary oxygen saturation was measured at 85% in all four limbs.

A two-dimensional transthoracic echocardiogram showed right ventricular hypertrophy, flattening of the interventricular septum, and normal biventricular function. Nevertheless, no congenital heart defect was found on conventional views (Panel A).

In order to further investigate the disease, a cardiac computed tomography (CT) scan was achieved. A huge connection between aorta and main pulmonary artery was beautifully observed (Panels B and C).

Aortopulmonary window is a rare congenital heart disease, due to failure of septation between the ascending aorta and the pulmonary artery during Foetal life but two separate semilunar valves are present. It produces a large left-to-right shunt, leading to congestive heart failure and, finally, Eisenmenger syndrome occurs at an early age because of combined systolic and diastolic run-off into the pulmonary circulation. Surgical correction is the treatment of choice in early stages of disease, but is contra indicated once the patient suffers from Eisenmenger syndrome. In the situation of extracardiac bidirectional shunt with low velocity such as APW or ductus arteriosus, echocardiography may be very challenging. Thus, if unknown, in case of cyanosis associated with pulmonary hypertension the site of the shunt can be reliably assessed by CT scan. (Panel A) Transthoracic echocardiography parasternal short-axis view above the level of valves showing normal cardiac structure and no evidence of congenital heart disease; (Panel B) CT angiography showing the aortopulmonary window with two distinct semilunar valves; (Panel C) Cinematic rendering reconstruction of CT angiography showing a massive APW.

Published on behalf of the European Society of Cardiology. All rights reserved. © The Author(s) 2018. For permissions, please email: journals.permissions@oup.com.