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Images in Congenital Cardiac Disease

Echocardiographic diagnosis of divided right atrium

Anke Schneider, Vít Rázek, Frank T. Riede

Department of Paediatric Cardiology, University of Leipzig, Leipzig, Germany

Keywords: Cor triatriatum dexter; hepatic cirrhosis; congenital heart disease

An 8-year-old girl presented at our institution with a 6 month history of increasing peripheral oedema, ascites, hepatosplenomegaly, and biopsy-proven hepatic cirrhosis of unknown aetiology. Transthoracic echocardiography revealed leftward deviation of the atrial septum, and dilation of the caval veins. Flow from the caval veins was restricted by a single opening in a shelf separating an anterior, supratricuspid, component of the right atrium from a posterior systemic venous sinus to which the caval veins connected (Fig. 1). The mean pressure gradient between the two atrial components, estimated by pulse wave Doppler, was 20 mmHg. Transoesophageal echocardiography confirmed the diagnosis of divided right atrium (Fig. 2; to see movie clips of echocardiography visit website http://journals.cambridge.org.cty). The dividing shelf was removed completely at cardiac surgery, with postoperative transthoracic echocardiography confirming its complete excision, and showing

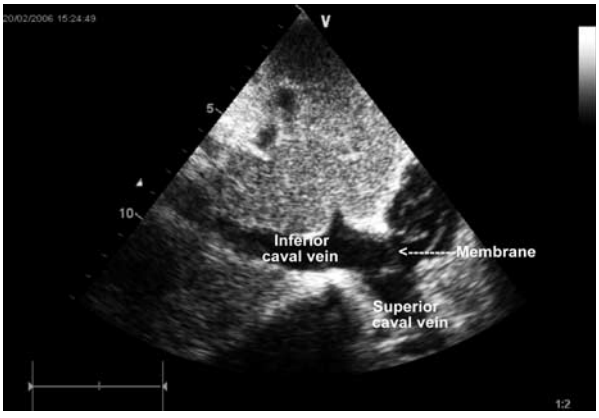

Figure 1.

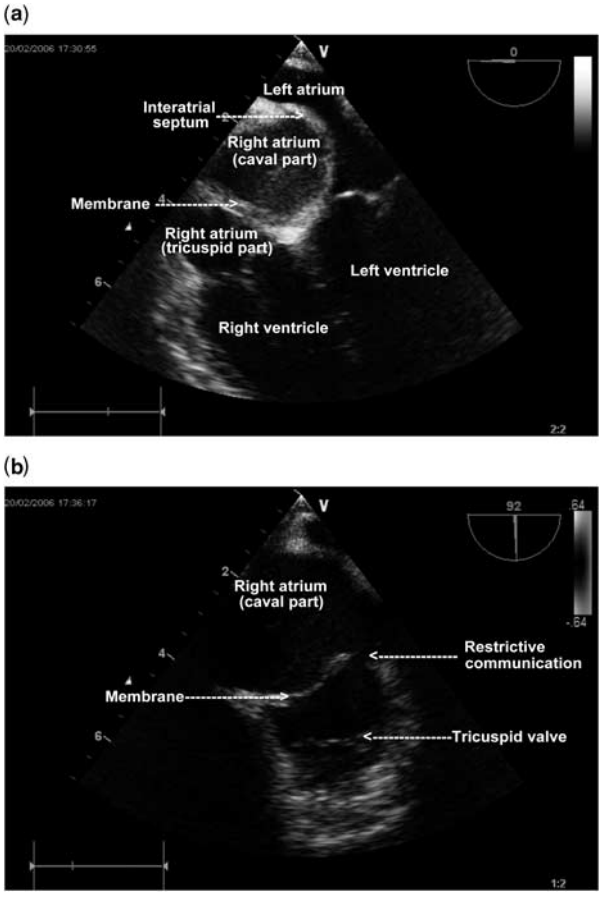

Figure 2.

Correspondence to: Anke Schneider, University of Leipzig, Dept. of Paediatric Cardiology, Strümpellstr. 39, 04289 Leipzig, Germany. Tel: +49 341 8561036; Fax: +49 341 8651143; E-mail: schmia@medizin.uni-leipzig.de; Anke.Schneider@med.uni-duesseldorf.de

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subsequent free flow from the systemic veins to the tricuspid valve. The postoperative course was uneventful and, at follow-up 1 year later, she remains free from symptoms. Hepatosplenomegaly has regressed, and hepatic function is now normal.

Division of the right atrium,[1] also known as cor triatriatum dexter, is a rare congenital abnormality in which persistence of the right valve of the embryonic systemic venous sinus divides the right atrium into two chambers. It may be associated with other congenital malformations, such as pulmonary stenosis or atresia, tricuspid atresia, and hypoplastic right ventricle. Clinical symptoms are highly variable, and depend on the degree of obstruction between the chambers. Our patient illustrates the potential for the dividing shelf to produce severe obstruction to systemic venous return, and the excellent results achieved by surgical resection.[1]

Reference

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