## **Persistent, Imperforate Eustachian Valve**

Tammo Delhaas, MD, PHD,\*† Marc Gewillig, MD, PHD,‡ Marcus Lance, MD,§ Cees Wittens, MD, PHD||

Maastricht, the Netherlands; and Leuven, Belgium



From the \*Department of Pediatrics, Cardiovascular Research Institute Maastricht, Maastricht University Medical Center, Maastricht, the Netherlands; †Department of Biomedical Engineering, Cardiovascular Research Institute Maastricht, Maastricht University Medical Center, Maastricht, the Netherlands; ‡Department of Pediatric Cardiology, University Hospitals Leuven, Leuven, Belgium; §Department of Anesthesiology, Cardiovascular Research Institute Maastricht, Maastricht University Medical Center, Maastricht, the Netherlands, and the ||Department of Surgery, Cardiovascular Research Institute Maastricht, Maastricht University Medical Center, Maastricht, the Netherlands. Manuscript received November 23, 2012; accepted November 27, 2012.

42-year-old man with a history of neurofibromatosis type 1 and recurrent right femoral vein thrombosis presented to the emergency department with inability to walk due to severely swollen legs. He took anticoagulants and was wearing compressive stockings. A computed tomographic scan of the chest and abdomen showed intrahepatic neurofibromata, a small inferior vena cava (IVC), and extensive paravertebral venous collaterals (A). Angiography (B) and agitated saline contrast echocardiography (C) with injection into the right femoral vein showed the suspected occlusion of the IVC to be at the junction of the IVC with the right atrium (RA) due to a persistent, imperforate Eustachian valve (EV). Antenatally, the EV directs blood from the IVC towards the foramen ovale. Postnatally, this valve regresses and is usually barely visible in adults.

To relieve obstruction, we performed transcatheter blade and balloon valvotomy (**D**), resulting in unobstructed flow from the IVC to the RA (**E**). The patient's condition started to improve 1 week after intervention. Upon discharge after 3 months, he could walk again. His weight had dropped from 162 to 134 kg, and his right and left upper leg circumferences had decreased from 92 and 82 cm to 74 and 67 cm, respectively.