

Dealing with an unroofed coronary sinus in a delicate clinical condition.

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Introduction:

Unroofed coronary sinus is an uncommon type of atrial septal defect in which a communication occurs between the coronary sinus and the left atrium as a consequence of an absent or deficient coronary sinus septum.

The diagnosis may be often overlooked because of the lack of specific clinical features and the difficulty to define the exact anatomy by transthoracic echocardiography since the defect is not always oval and the coronary sinus is not often a perfect cylinder.

Methods and Results:

We describe a 17 year-old girl with hepatic failure that was assessed by a pediatric cardiology team before a liver transplant. She was affected of a portal cavernomatosis initially treated with a transjugular intrahepatic portosystemic shunt (TIPS) due to portal hypertension. Transthoracic echocardiography showed a right ventricular dilatation. Also, an unroofed coronary sinus (UCS) was suspected. No persistent left superior vena cava was observed. Magnetic resonance imaging confirmed a complete absence of the roof of the coronary sinus with an enlarged ostium (asterisk in the figure), and a Qp/Qs ratio of 1.9. In a multidisciplinary meeting a percutaneous approach to close the coronary sinus atrial septal defect type was decided given the delicate condition of the patient. Transesophageal echocardiography helped to define anatomy (Figure 1A) and to guide device position. The right femoral vein was cannulated and an angiogram from the left atrium demonstrated the coronary sinus atrial septal defect (Figure 1B). A Figulla Flex II ASD occluder™ 27mm was successfully deployed. There was no residual shunt or atrio-ventricular valves interference, and the patient remained on sinus rhythm at all times. The device stopped the left to right transatrial shunt, leaving the unroofed coronary sinus draining to the left atrium. The right sided chambers dilatation normalized progressively, and six months later the device remains stable in a normal position.

Conclusions:

Although surgery is still the mainstay for unroofed coronary sinus closure, transcatheter-based management is quickly evolving and it may represent a valid, safe and less invasive alternative to conventional heart surgery.

