

Persistent Left Superior Vena Cava Closure In a Patient With Unroofed Coronary Sinus

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Introduction: Unroofed coronary sinus (URCS) is a rare congenital cardiac anomaly and has been almost always associated with a persistent left superior vena cava (PLSVC). PLSVC draining into the left atrium is responsible for a right-to-left shunt that can be a source of oxygen desaturation. Systemic thromboembolism and brain abscesses are possible complications of this condition. In order to prevent recurrent cerebral emboli and abscesses, correction of this anomaly has been recommended. Although transcatheter based treatment therapies are used to correct this condition in adults, there was a little experience in children.

Case Report: A 6 year-old girl was admitted with cough and fever. Physical examination revealed slight cyanosis and clubbing of the fingers. Transthoracic echocardiographic evaluation with agitated saline injection from the left arm revealed a PLSVC draining into the left atrium suggesting URCS. Diagnosis was confirmed by CT. Under general anesthesia, a catheterisation was performed with left jugular venous and right femoral venous access. An angiogram through the left jugular vein confirmed the LSVC draining directly into the left atrium. There was no significant pressure rise in LSVC during test occlusion of the distal part of the LSVC. Occlusion angiography showed sufficient drainage to the right SVC via bridging vein. Sheath advanced through the coronary sinus to the LSVC. Distal part of the LSVC was occluded with 16/12 mm Amplatzer Vascular Plug II[®] (below the drainage of the accessory hemiazygos vein). Control angiography demonstrated that there was no residual shunt (Figure). After the procedure, arterial oxygen saturation in room air immediately increased from 91% to 98%. Patient was discharged the day after the procedure without any complication.

Conclusion: If URCS left untreated it can cause complications such as systemic thromboembolism, infective endocarditis, and brain abscess. Early diagnosis and treatment are very important. Although surgery has good results, in selected cases, transcatheter closure appears to be a safe and effective alternative to surgical treatment.



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