

WCC 2016-113 : Asymptomatic presentation of severe aortic stenosis with unicuspid aortic valve: Madhuri Sabinkar, Madhuri Sabinkar, Y.V.Subba Reddy, O. Adikesava Naidu, Ravi Srinivas, Srikanth Eruvaram

Introduction : The estimated incidence of congenital unicuspid aortic valve in the adult echocardiographic population was reported as about 0.02%. It was often discovered during surgery for aortic stenosis or post mortem.

The unicuspid aortic valve is an extremely rare congenital anomaly. It usually presents with aortic stenosis and/or aortic regurgitation. Other cardiovascular complications, such as aortic dilatation and left ventricular hypertrophy can accompany it.

Unicuspid aortic valve shares many of the features of bicuspid aortic valve, including valvular dysfunction, aortic dilatation, aortic dissection, and dystrophic calcification, although these conditions develop at an earlier age and progress at a faster pace in unicuspid aortic valve.

Case report :

An asymptomatic 8 year old girl was incidentally detected to have a murmur during a medical camp conducted at school and was referred to our hospital for evaluation. Physical examination revealed cardiomegaly, heaving apical impulse, soft aortic component of second heart sound and an ejection systolic murmur in aortic area radiating to apex and carotids. Trans thoracic echocardiography revealed unicuspid aortic valve with severe aortic stenosis.

Discussion :

Unicuspid aortic valve presenting at this age is rare and is often confused with a bicuspid valve, a more common congenital abnormality. In adults undergoing surgical intervention for aortic valve disease, unicuspid valve accounts for $\leq 5\%$ of the cases.