Intermediate type of Gerbode defect: rare type of the left to right shunt

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DESCRIPTION
Gerbode defect is the shunt originating from the left ventricle (LV) and going into the right atrium (RA) during systole. Three types of defect have been reported in the literature: type 1 (indirect or infravalvular defect in the interventricular part of the membranous septum), type 2 (direct or supravalvular defect in the atroioventricular (AV) part of the membranous septum) and type 3 (intermediate, in the both AV and interventricular part).

We report a case of the 2-year-old female child presented with the frequent chest infections and failure to thrive. She had grade 4/6 pansystolic murmur at the left parasternal area and wide split with normal intensity second heart sound. On the X-ray pulmonary vascularity was increased and the RA was enlarged. On echocardiography, there is a ventricular septal defect (VSD) of size 5 mm with L-R shunt with a gradient of 77 mm Hg (figure 1). On apical four-chamber view, there is VSD with colour jet noticed simultaneously from an LV to the right ventricle (RV) and from the tricuspid valve (TV) to the RA (video 1, figure 2).

On parasternal short axis, simultaneous LV-RV and TV-RA shunt are demonstrated (figure 3). Initially, the diagnosis of indirect (infravalvular) type of Gerbode defect was made. But on subcostal four-chamber view, LV to RA shunt is noted above the TV (supravalvular) (video 2, figure 4). So final diagnosis of intermediate (type 3) Gerbode defect was made. The child underwent cardiac catheterisation and was sent for the surgical correction.

Gerbode defect is an extremely rare anomaly as only six cases had been diagnosed in children memorial hospital in Chicago during the 18-year period. It can be congenital or acquired after valve surgeries, correction of shunt defects and rarely seen in infective endocarditis or after AV node ablation. The LV to RA shunt can occur at three levels during systole:

Type 1 (indirect type): In a perimembranous VSD, the shunt originates below TV from a LV to RV and simultaneously through the TV into the RA. The TV may have a cleft, widened commissural space, perforation, abnormal chordate and other deformities.

Type 2 (direct type): In a true or direct Gerbode, the blood in the LV goes through the small area of

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Video 1 AP4C (apical 4 chamber) view showing infravalvular type of Gerbode defect. 2D, two dimensions.

Video 2 Subcostal four-chamber showing supravalvular type of Gerbode defect. 2D, two dimensions.

the membranous septum above the TV into the RA. It is rare than indirect.5

Type 3 (intermediate): This is combination of direct and indirect type.

X-ray shows an enlargement of the RA and increased pulmonary flow. Echocardiography is the mainstay of the diagnosis and helpful in differentiating this entity from the ruptured sinus of Valsalva into the RA (continuous shunt), endocardial cushion defect, VSD with TR. There is a significant gradient from LV to RA shunt on Doppler interrogation. Sometimes false interpretation of pulmonary artery hypertension (PAH) can be made. A normal diastolic pulmonary arterial pressure estimated from the pulmonic regurgitation jet is helpful for distinguishing true PAH from the high-velocity jet in the RA caused by Gerbode defect. Silbiger et al1 specified several key echocardiographic clues suggesting Gerbode defect, including1: atypical jet direction3;

shunt during systole2; lack of ventricular septal flattening3; no right ventricular hypertrophy and3 normal diastolic pulmonary artery pressure as estimated from the pulmonic regurgitant velocity. The diagnosis can be confirmed on TEE, cardiac CT and cardiac catheterisation.

Learning points

► In case of suspicion of ventricular septal defect on clinical examination, enlargement of the right atrium on X-ray, the Gerbode defect should be ruled out.
► Sometimes Gerbode defect can be misinterpreted as pulmonary artery hypertension, in this scenario absence of right ventricular hypertrophy, lack of septal flattening and normal pulmonary artery diastolic pressure favours the diagnosis of Gerbode defect.

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