Apical ventricular septal defect with reverse bidirectional transseptal flow: a sign of restriction within the right ventricle

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Apical ventricular septal defects (VSD) may spontaneously become restricted or may even close following tissue outgrowth within the right ventricle in the region of the apical muscle bundles. We report a case of spontaneous restriction of an apical VSD localized within the right ventricle in the region of prominent apical trabeculae, in which pulsed Doppler interrogation showed an unusual pattern of bidirectional blood flow across the VSD, with right to left flow in systole and left to right in diastole. Angiography confirmed these unusual aspects. This reverse bidirectional transseptal flow is due to the physiological incorporation of the apical part of the right ventricular cavity in the left ventricle and may be, in the absence of right outflow obstruction, a sign of restriction of the VSD inside the right ventricle.

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Bidirectional blood flow across moderate and large ventricular septal defects (VSDs) has been demonstrated at cardiac catheterization and angiography[1] and Doppler echocardiography[2,3]. In the absence of an elevated pulmonary resistance or of right outflow obstruction, a left to right transseptal flow in systole and a right to left flow in early diastole, with reversed flow in late diastole, are usually observed. Conversely, in small VSDs a continuous left to right shunt is observed throughout the cardiac cycle. We report a case of a moderately sized muscular VSD with an unusual pattern of reverse bidirectional transseptal flow. Echo-Doppler study and angiography showed that restriction of the left to right shunt occurred inside the right ventricle.

Case report

A child, aged 8 years, with clinical signs of a small ventricular septal defect underwent two-dimensional and echo-Doppler study. A cross-sectional, two-dimensional echocardiography was performed in the apical 4-chamber view and parasternal long- and short-axis views to demonstrate the VSD location, anatomy and size. The subxyphoid 4-chamber and transverse views were also obtained. The two-dimensional echoes clearly demonstrated an apical VSD infero-posterior to the moderator band and 13 mm in diameter (Fig. 1). Pulsed Doppler study of the transseptal flow was performed. The transducer was placed to the left of the cardiac apex so as to pass the cursor line in a plane almost perpendicular to the apical interventricular septum and then the sampling volume was positioned in the center and on the right side of the defect. In this left lateral position, the Doppler signals towards the transducer (positive deflection) indicate right to left flow, and Doppler signals away from the transducer (negative deflection) indicate left to right flow. When the sampling volume was positioned within the defect, a to and fro low velocity flow, right to left in systole and left to right in diastole, was observed (Fig. 2A). This transseptal Doppler flow pattern is almost the opposite of the flows usually observed in case of VSDs. Color flow mapping and Doppler interrogation showed that the shunt flow inside the right ventricular chamber was restricted between the septomarginal and minor apical trabeculae (Figs. 2B and 2C). At cardiac catheterization, the patient was found to have a small left to right shunt at the level of the ventricles, on the basis of the oxygen saturation. Angiography confirmed the anatomy of the VSD and the right intraventricular mechanism of restriction (Fig. 3). It was a restrictive VSD with a pulmonary to systemic flow ratio of 1.3 and with normal ventricular pressures.

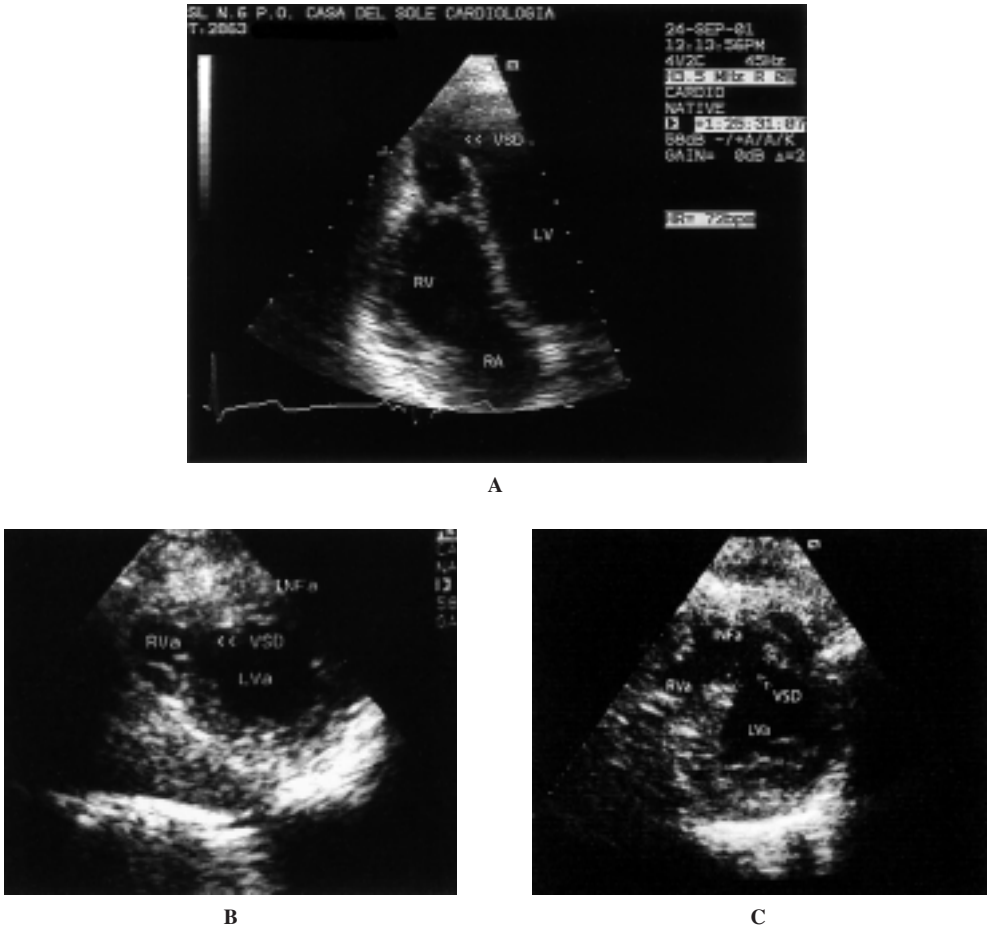
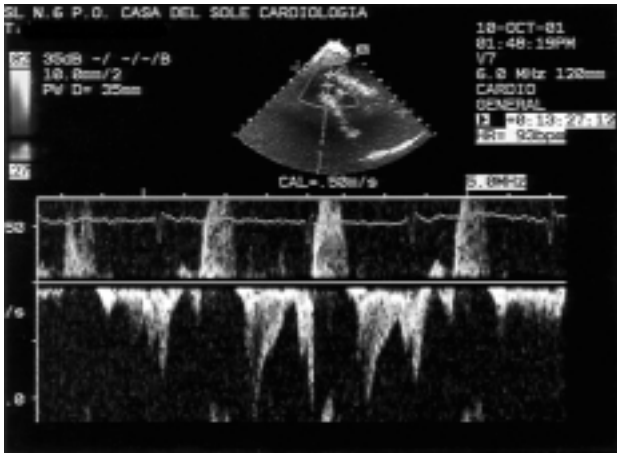


Figure 1. *Transthoracic echocardiographic imaging of the apical ventricular septal defect (VSD)* (arrowheads). A: *apical 4-chamber view demonstrating the muscular VSD distal to the septomarginal trabecula of the right ventricle (RV).* B: *magnified parasternal apical short-axis view showing the location of the VSD posterior to and to the right of the septomarginal trabecula, between the left ventricular apex (LVa) and the right ventricular sinus apex (RVa), while the infundibular apex (INFa) is anterior to and to the left.* C: *for comparison a parasternal short-axis view of a VSD located anterior to and to the left of the moderator band between the LVa and the INFa. LV = left ventricle; RA = right atrium.*

Discussion

Apical muscular VSDs, often multiple and with oblique channels and more openings on the right ventricular than on the left ventricular side, are located close to the heart apex, distal to the insertion of the moderator band. In the parasternal or subxyphoid short-axis views these defects may be distinguished in two subtypes: antero-superior to and to the left of the moderator band, and postero-inferior to and to the right of the moderator band. These subtypes have been regarded as mere topographic variants of the defects between the left and right ventricular apices[4,5] or as two anatomically distinct types[6-8]: the apical defects located in the anterior and left aspects of the apical septum represent communications between the left ventricular apex and the infundibular apex, that is the lowermost part of the right ventricular outflow tract. The apical VSDs located in the posterior and right sides are considered as defects between the left ventricular apex and the right ventricular sinus apex, that is the most apical part of the right ventricle inflow tract. Based on this distinction our case represents a defect between the left ventricular apex and the right ventricular sinus apex as clearly shown in figure 1. It is well known that, similar to all the muscular VSDs, many of these defects close spontaneously[9], although it has been demonstrated that the probability of closure is lower than for other muscular defects[10,11]. While the mechanisms of closure of perimembranous VSDs are well documented[12,13], the morphologic changes associated with spontaneous closure of the muscular VSDs have not been well understood and may be various. Some trabecular muscular defects appear to close by progressive growth of tissue from the right ventricular side[11,14]. Spontaneous closure of an apical VSD within the right ventricle at the level of the moderator band, resulting in the physiological incorporation of the infundibular apex within the left ventricle, was reported by Kumar et al.[6]. These authors also performed transcatheter closure of apical VSDs by placing the device inside the right ventricle at the narrowest part of the defect straddling the apical right trabeculae. The echocardiographic and angiographic images of our case showed that, similar to what observed



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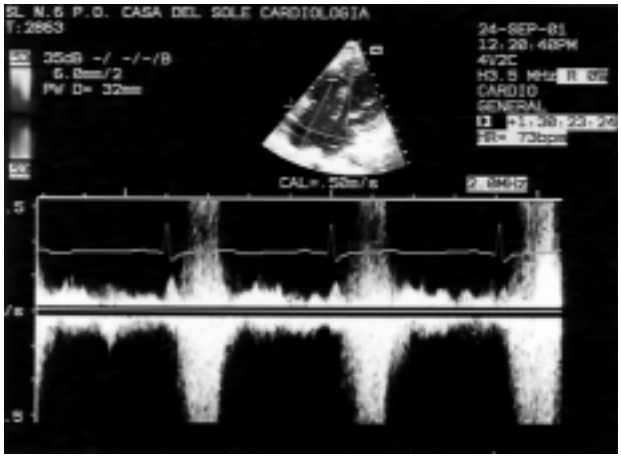

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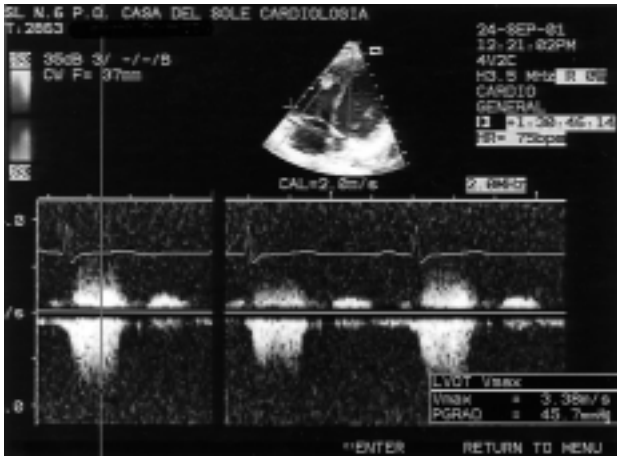
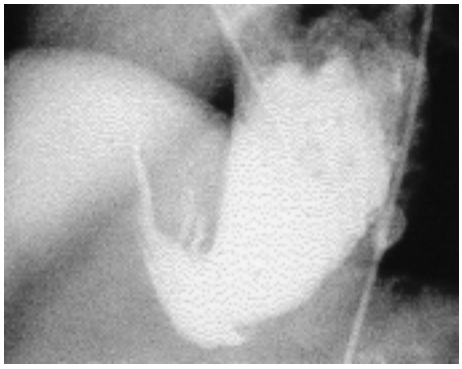


C

Figure 2. *Echo-Doppler analysis.* A: *electrocardiographic trace and pulsed echo-Doppler. The transducer was placed to the left of the cardiac apex and the sample volume to the right side of the ventricular septal defect. Owing to this left lateral position the Doppler signals towards the transducer (positive deflection) indicate right to left flow, and those away from the transducer (negative deflection) indicate left to right flow. In systole, mainly during end-systole, the flow is directed from the right to the left ventricle, while in diastole a biphasic left to right flow is evident. The signal variability is related to respiratory movements.* B: *the pulsed Doppler interrogation from the apical position. The sample volume was placed inside the right ventricular cavity at the level of the septomarginal trabecula and shows systolic, high velocity, disturbed flow.* C: *continuous Doppler showing the shunt flow away from the apical region of the right ventricle with a peak velocity of 3.38 m/s.*



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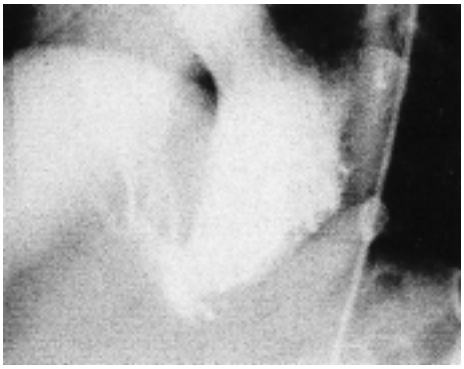

B

Figure 3. *Angiogram obtained during contrast injection in the left ventricle using a pigtail catheter.* A: *diastolic frame of the hepato-clavicular projection showing a large communication between the left ventricle and the apical region of the right ventricle.* B: *systolic frame showing contraction of the apical region of the right ventricle and a small restrictive communication with the rest of the right ventricle.*

by Van Praagh et al.[8], the region of prominent muscle bundles separating the right ventricular sinus apex from the rest of the right ventricular cavity was narrower than the defect in the apical septum. In this region within the right ventricle we observed the typical flow pattern of a restrictive VSD (high velocity, disturbed, "left to right" flow). Shifting the sample volume of the pulsed Doppler to the right side or to the center of the real VSD, a bidirectional flow with an unusual pattern became evident.

As previously described in angiography[1] or echo-Doppler studies[2,3], in case of VSDs transseptal shunting is a complex process. While the systolic flow direction and size depend on the resistance to flow in the left and right ventricular outflow tracts, diastolic flow occurring across the VSD depends on the differential rates of left and right ventricular contraction and relaxation. In case of moderate or large VSDs without an elevated pulmonary resistance or right outflow obstruction, the systolic flow direction is left to right, while the direction of the initial diastolic flow, during isovolumetric relaxation, is right to left. In the presence of right ventricular volume overload the flow across the VSD may be from the right to the left ventricle throughout diastole.

This bidirectional flow pattern reflects the intrinsic functional properties of the morphologic left and right ventricles: it is not related to ventriculo-arterial connections and it is not affected by the type of ventricular septal defect[3]. Our case showed a reverse bidirectional transseptal flow: during systole, mainly during end-systole, the direction of flow was right to left, and during the whole diastole there was a biphasic left to right flow. As the systemic oxygen saturation was normal, we presume that a small volume of blood was shunted from the right to the left ventricle during systole and returned to the right ventricle during diastole; thus, it did not enter the systemic circulation. This reverse bidirectional flow pattern is similar to flows encountered in case of ventricular diverticula[15]. We think that these atypical flows depend on the peculiar mechanism of restriction of some muscular VSDs which leads to the physiological incorporation of the apical part of the right ventricular cavity in the left ventricle.

In conclusion, apical VSDs may spontaneously become restrictive or may even close following tissue outgrowth within the right ventricle in the region of the apical muscle bundles. This particular mechanism of restriction is revealed by a peculiar flow pattern with reverse bidirectional flow across the VSD.

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