A 13-year-old boy was admitted for cardiac catheterization because of mitral regurgitation. The family history was noncontributory. He is the second twin, delivered after normal pregnancy without maternal illness, infection, or medication. He showed normal development and weight gain. At the age of 2 years, a heart murmur was first noted; echocardiographically, a small (2.5-mm) ventricular septal defect (VSD) was found. This closed spontaneously 3 years later, but mitral valve prolapse accompanied by mild mitral incompetence was noticed. During the next 2 years, mitral regurgitation became more severe. At the age of 10 years, mitral insufficiency was treated surgically because of the clinical signs of congestive heart failure. Afterwards, mild mitral regurgitation was still noticed in echocardiography. Clinically, the boy did well during the next 3 years, but at echocardiography, the left atrium as well as the left ventricle were found to be progressively enlarged, probably caused by mitral regurgitation. The boy again was admitted to the hospital for catheterization.

We saw a well-nourished 13-year-old boy without hyperactive precordium and normal pulses. The first heart sound was loud; a holosystolic heart murmur grade 3/6, best heard at apex, was found. The ECG showed sinus rhythm, 92 beats/min with normal intervals; T-wave depression in V1-V3; and bundle-branch block configuration in lead 1. Chest radiograph showed an atypical cardiac silhouette with a round calcification at the left cardiac border (Fig 1). This could be detected in exactly the same size and locus on older radiographs for > 9 years. Cardiac catheterization showed, beside the mitral regurgitation, an outpouching of the free left ventricular wall with a round calcification at the top. This outpouching had a narrow connection to the ventricular cavity, which was contracting simultaneously with the ventricle (Fig 2). The injection into the aortic root showed normal coronary arteries without connection to the outpouching. A former catheterization 4 years ago showed exactly the same outpouching.

What is the diagnosis?
Figure 1. Chest radiograph showing a round calcification at the left cardiac border (arrow). Seven years ago, this finding was identical.

Figure 2. Left ventricular angiogram showing the diverticulum with narrow connection to the ventricle and a round calcification at the top (arrows).
Diverticula of the ventricle are sometimes discovered accidentally even in adults. It seems to be a rare malformation: in 750 necropsies, 0 to 4% had this malformation; only 10 of 13,000 congenital heart disease operations were performed for this reason. The first description by O’Bryan is dated 1836. Since then, only about 150 left ventricular diverticula, either isolated or in association with midline defects, were reported. The largest series of nine patients with true congenital ventricular diverticula was published by Hamaoka et al in 1987, who examined about 2,000 children. Bharati et al found nine right ventricular diverticula in > 3,000 necropsies.

The history of our patient gives no clue for previous infection, trauma, or infarction; therefore, a congenital malformation is probable. The association with mitral incompetence has been described before, but most of the known diverticula with this association have their location more subvalvular than in our patient. Neither the papillary muscles nor the coronary arteries are involved. It is most likely an unassociated finding. The calcification could be explained by the narrow connection with restriction to flow. The cause of the mitral incompetence is unclear, as in the two cases reported by Gueron et al, who tried to classify outpouchings of either ventricle for the first time by clinical and angiographic features.

The etiology of cardiac diverticula remains uncertain and has been subject of speculation. For Cantrell’s syndrome with apical diverticula associated with midline thoracoabdominal defects, a failure of the normal midline fusion of the paired primitive mesoderm, probably with abnormal fusion of the cardiac loop to the yolk sac before its descent, is suggested. The fibrous type might be caused by a focal weakening of the ventricular wall caused either by an intrinsic abnormality during embryogenesis or acquired in utero by virus infection. The ventricular pressure may produce a diverticulum in this area of abnormal weak myocardium. It remains unclear why diverticula then are able to contract simultaneously with the ventricle after birth.

There are reports of progression of fibrosis and of the lesions by age and time. As in the case reported by Archbold et al, the diverticulum did not change in size over for several years, as retrospectively noted. Because the history of our patient gave no clues for complications (like arrhythmia or thrombus formation) and the course showed no progression, the diverticulum was left untreated.
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