Congenital cardiac diverticulum in the subaortic valve area

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Objective: We report and review the clinical characteristics and operative techniques of the rare malformation of congenital cardiac diverticulum in the subaortic valve area.

Methods: Four male patients with congenital ventricular fibrous diverticula in the subaortic valve area are reported.

Results: The patients' ages ranged from 4 to 47 years (mean 39 years). The first case was complicated by rupture, pseudoaneurysm formation, and a hemopericardium, and was only diagnosed on the third surgical exploration. The second case was associated with endocarditis involving both aortic and mitral regurgitation. The diverticulum was suspected on 2-dimensional transthoracic echocardiography and confirmed at surgery. In the third case, the subaortic diverticulum was associated with a rupture of the left cusp of the aortic valve with severe regurgitation. In the fourth case, the diverticulum was located between the aortic and mitral valves with a diameter of approximately 30 mm and was associated with multiple perforations of an aortic valve leaflet and severe regurgitation. There were no other congenital thoracoabdominal or cardiac anomalies. At a mean follow-up of 1 year, all patients were well.

Conclusions: Congenital cardiac diverticulum in the subaortic valve area is a rare anomaly. Transthoracic echocardiography and transesophageal echocardiography are reliable tools for the diagnosis and follow-up. Isolated congenital cardiac diverticula in the subaortic valve area tend to involve a benign clinical process, but may be complicated by spontaneous rupture or endocarditis. Surgical resection is performed when complications require surgical therapy.

Cardiac diverticulum is a rare congenital malformation, with approximately 100 cases reported in the English literature. It was reported in only 0.4% of 750 autopsy cases (3 cases) with cardiac death,¹ and in 0.076% of more than 13,000 congenital heart operations (10 cases).² Most diverticula arise as an out-pouching from the left ventricle, or more rarely from the right ventricle,^{3,4} or both.^{5,6} They are usually associated with other cardiac anomalies and sometimes part of the congenital midline thoracoabdominal malformation syndrome.⁷ Some authors have advocated resection of all diverticula to prevent rupture,^{2,8} whereas others have adopted conservative management of asymptomatic isolated diverticula.^{9,10}

This report presents 4 male patients (age range 4–47 years, mean 39 years) with a subaortic diverticulum complicated by the rupture or prolapse of an aortic valve cusp and severe regurgitation. Pulsatile paradoxical accessory chambers of the left ventricular outflow tract were the typical appearance on 2-dimensional echocardiography, suggesting that there was no myocardium within the diverticula. All patients underwent exploratory cardiac surgery confirming the diagnosis. Histopathology demonstrated fibrous ventricular diverticula.

Case Reports

Case One

A 4-year-old boy was referred to another hospital after sudden syncope. Physical examination revealed a pale face, a rapid and weak pulse, a heart rate of 140

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Abbreviations and Acronyms

CPB = cardiopulmonary bypass

TTE = transthoracic echocardiography

beats/min, a blood pressure of 60/30 mm Hg, an SpO₂ of 94%, and jugular venous distension. Both pupils were equal and reactive to light. Computed tomography scanning of the chest and echocardiography both suggested a pericardial effusion and cardiac tumor. At exploration through a median sternotomy there was a moderate hemopericardium, but no cardiac tumor, bleeding site, or other abnormality was identified. The hemopericardium was evacuated, and the chest was closed.

Fourteen days later, the boy was referred to our institute for further evaluation of cardiac performance. Examination results were unremarkable. Specifically, there were no abdominal wall defects, abnormal precordial pulsations, or murmurs. Transthoracic echocardiography (TTE) revealed good overall left ventricular systolic function and normal cardiac development and morphology, with the exception of a tongue-like accessory chamber between the aortic and mitral valves that connected to the left ventricular outflow tract through an 18-mm diameter ostium. The chamber was approximately 32 mm in length and 21 mm in width, increasing during systole and reducing during diastole (Figure 1, A, B). This classic paradoxical motion was considered characteristic of a fibrous diverticulum. No other significant intracardiac congenital anomalies were found.

At reexploration 31 days later after the first operation, a 40 × 30-mm pulsatile mass located between the aortic root and superior vena cava was found. On cardiopulmonary bypass (CPB), the mass was opened through its thin-walled (0.5 mm) apex and 2 orifices were found, one 6 mm and one 3 mm in diameter. A pericardial patch was used to close the orifices. On day 6 postoperatively, a TTE revealed a similar image of the accessory chamber as preoperatively. On day 12 the chest was reopened, and the diverticulum was found between the ascending aorta and superior vena cava, approximately 30 × 20 mm in diameter (Figure 1, C), with a 20-mm orifice freely communicating with the left ventricular outflow tract (Figure 1, D). A polytetrafluoroethylene (Gore-Tex; Flagstaff, Ariz) patch was used to close the orifice of the diverticulum. The surgery was uneventful, and the boy recovered. TTE examination confirmed the absence of the chamber. Histopathology demonstrated a fibrous diverticulum. At 1-year follow-up, the boy remained well.

Case Two

A 30-year-old man was referred to our institute with intermittent fever, fatigue, indigestion, and weight loss during the previous 4 months. His fever was controlled with antibiotic therapy. His medical history included acute nephritis

Figure 1. Case 1. TTE revealed a tongue-like accessory chamber between the aortic and mitral valves (A) that communicated with the left ventricular outflow tract through an 18-mm diameter ostium (B). The chamber was approximately 32 mm in length and 21 mm in width. It increased during systole and reduced during diastole. Day 12 after the second surgery, the chest was reopened, and a diverticulum was found between the ascending aorta and superior vena cava, approximately 30 × 20 mm in diameter (C). During the third surgical procedure, a diverticulum was found between the ascending aorta and superior vena cava, with a 20-mm orifice freely communicating with the left ventricle (D). LV, Left ventricle; LA, left atrium; AO, aorta. Diverticulum (arrow).

and hepatitis with jaundice. Physical examination showed normal body habitus, vital signs, and cardiac rhythm. Cardiomegaly was observed, a diastolic murmur was heard at the left sternal edge, and a 3/6 grade systolic murmur was heard at the apex. Pulmonary and abdominal examination results were unremarkable.

TTE revealed a bicuspid aortic valve with thickened leaflets and increased echo-reflection, multiple perforations, and granular or striplike vegetations attached to the valve. The left coronary cusp exhibited moderate prolapse into the left ventricular outflow tract, and color-flow Doppler confirmed severe aortic regurgitation with a regurgitant flow speed of 4.3 ms and pressure gradient of 74 mm Hg. The mitral leaflets were thickened with granular vegetations attached to them and their chordae. There was moderate mitral regurgitation. A 30-mm diameter semi-spherical accessory chamber arose through a 13-mm orifice from the wall of the left ventricular tract beneath the aortic valve. The left ventricle was dilated with an end-systolic diameter of 65 mm. A diagnosis of congenital fibrous left ventricular sub-aortic diverticulum with endocarditis and aortic/mitral regurgitation was made, and surgery was recommended.

At sternotomy on CPB, a 30-mm diameter pulsatile mass was found between the aortic root and superior vena cava

Figure 2. Case 2. During the surgical procedure, a 30-mm diameter pulsatile cyst was found between the aortic root and superior vena cava (C, this image was taken after closing the ostium of the diverticulum, so it appears smaller than it was). The ostium of the accessory chamber, which was 20 mm in diameter, was mainly located below the left coronary sinus with intact endothelium (A). The ostium of the diverticulum was closed with a Dacron patch through the aortic incision (B). The diverticulum was resected outside of the aorta (D).

(Figure 2, C). A bicuspid aortic valve with multiple vegetations and 2 perforations of the left coronary cusp was confirmed. The mitral valve leaflets were thickened with multiple vegetations attached. The ostium of the accessory chamber was 20 mm in diameter and located beneath the left coronary cusp; the chamber was lined with endothelium (Figure 2, A). The ostium of the diverticulum was closed with a Dacron patch through the aortic incision (Figure 2, B), and its cyst was resected outside of the aorta and covered with a pericardial patch (Figure 2, D). The aortic and mitral valves were replaced with bileaflet mechanical prostheses. Histopathology demonstrated a fibrous diverticulum. Postoperative recovery was uneventful, and echocardiography on day 6 showed an intact repair of the ostium of the diverticulum and good prosthetic valve function.

Case Three

A 16-year-old male patient had experienced intermittent palpitations after active excise during the previous 4 years. On physical examination, cardiac rhythm was normal. There was cardiomegaly with a diastolic murmur heard at the left sternal edge. There was a wide pulse pressure and water-hammer pulse. Pulmonary and abdominal examination results were unremarkable.

TTE demonstrated prolapse of the left coronary cusp of the aortic valve into the left ventricular outflow tract, with a 0.3-cm perforation. The left ventricle was enlarged with an

Figure 3. Case 3: The diverticulum was located behind the left ventricular outflow tract (A). Color-flow Doppler revealed pulsatile flow into the chamber during systole (B). Case 4: During the surgical procedure, a 30-mm diameter pulsatile cyst was found between the aortic root and superior vena cava (C). The ostium of the accessory chamber, which was 20 mm in diameter, was mainly located below the left coronary sinus with intact endothelium (D).

end-diastolic diameter of 73 mm. A 40 × 38-mm diameter semi-spherical accessory chamber arose through a 21-mm orifice from the wall of the left ventricular outflow tract (Figure 3, A). There was moderate to severe aortic regurgitation with a regurgitant flow speed of 4.9 ms, pressure gradient of 95 mm Hg, and pulsatile flow into the accessory chamber during systole (Figure 3, B). A diagnosis of congenital fibrous left ventricular subaortic diverticulum with severe aortic regurgitation was made, and surgery was recommended.

At sternotomy with CPB, a 40 × 38-mm pulsatile mass was found between the aortic root and superior vena cava with the orifice below the left coronary sinus. There was a perforation through the left cusp of the aortic valve, which was thickened with rolled leaflet edges and severe regurgitation. The ostium of the accessory chamber was 20 mm in diameter and was closed with a 4-0 polypropylene continuous suture. The aortic valve was replaced with a bileaflet mechanical prosthesis. At follow-up, echocardiography showed a clotted accessory chamber and good prosthetic function.

Case Four

A 47-year-old man was referred to our hospital with intermittent fever and nasal obstruction. Physical examination revealed a blood pressure of 131/63 mm Hg, cardiomegaly, and a diastolic murmur at the left sternal edge.

TTE revealed an aortic valve with thickened leaflets, increased echo-reflection, and granular or striplike vegetations. The right coronary cusp prolapsed into the left ventricular outflow tract with a 4-mm regurgitant orifice. A 25 × 6-mm diameter semi-spherical accessory chamber arose through a 4-mm orifice from the left ventricular outflow tract. The chamber was pulsatile with movement paradoxical to the ventricle. A diagnosis of congenital fibrous left ventricular subaortic diverticulum and partial aortic valve prolapse with severe regurgitation was made.

At sternotomy with CPB, a 30-mm diameter pulsatile mass was found between the aortic root and superior vena cava (Figure 3, C). The aortic leaflets were perforated in multiple sites, and multiple vegetations were attached. The accessory chamber orifice beneath the left coronary sinus (Figure 3, D) was closed with a Dacron patch, and the valve was replaced with a bileaflet mechanical prosthesis. The patient recovered well.

Discussion

Congenital cardiac diverticulum is a rare congenital anomaly. Since it was first described by O'Bryan in 1838,¹¹ there have been approximately 100 cases reported in the English literature with a prevalence estimated at 0.013%.¹² In the English literature, most congenital cardiac diverticula were found in newborns and children, but involving all age groups from as young as 4 hours¹³ to 81 years.¹⁴ It has also been detected at fetal echocardiographic examination.^{15,16} Most cardiac diverticula arise from the left ventricle, but right ventricular,^{3,4} right atrial,¹⁷ left atrial,^{6,18} and dual ventricular origins^{5,6} have been reported, single or multiple.^{5,6,19} The cardiac diverticulum arose from just beneath the aortic valve in each of our patients.

The most frequently described pathologic classification is muscular and fibrous types,²⁰ but rarely may be mixed.²¹ Pathologic examination of muscular diverticula reveals all 3 cardiac layers within its wall representing true diverticula. They usually arise from the left ventricular apex by a narrow neck. They are frequently associated with midline thoracoabdominal defects (Cantrell's syndrome) or various other congenital cardiac malformations.¹³ For this reason, they are discovered more frequently in infancy. Vazquez-Jimenez and associates,¹³ in a well-conducted literature review in 1998, described 153 cases of Cantrell's syndrome, including left ventricular diverticula in 41 patients (32.3%) and right ventricular diverticula in 4 patients (3.1%).

The fibrous type is found near the base of the atrioventricular valve ring or at the apex. The fibrous type is not associated with midline defects of congenital cardiac malformation. Histology reveals predominantly fibrous tissues. Most fibrous diverticula are clinically silent.

The cause of cardiac diverticula is uncertain. The association between apical muscular diverticula and Cantrell's syn-

drome suggests a common defect in embryologic development for this abnormality. A local weakening in the developing ventricular muscle may give rise to other diverticula.²²

The clinical representation of congenital cardiac diverticula varies, from asymptomatic or atypical chest pain to severe congestive heart failure or sudden death, mainly depending on the associated cardiac abnormalities. Congenital cardiac diverticula may be asymptomatic until adulthood and are usually discovered incidentally during the assessment of an abnormal cardiac murmur by physical examination or other cardiac-related symptoms by using echocardiography or cardiac catheterization.

Echocardiography is a useful diagnostic tool to detect congenital cardiac diverticula. It allows an accurate assessment of morphology, location, and possible thrombosis in the diverticulum, and demonstrates other congenital cardiac abnormalities. Transesophageal echocardiography enhances the imaging definition and more clearly reveals the rear structure of the heart, which is helpful in diagnosing an inferior ventricular diverticulum.²⁶ Pulsatile paradoxical accessory chambers of the left ventricular outflow tract are the typical appearance on 2-dimensional echocardiography of congenital ventricular fibrous diverticula in the subaortic valve area, suggesting no myocardium within the diverticula. The congenital muscular ventricular diverticulum in 2-dimensional echocardiography appears as a tongue-like or digitiform outpouching of the ventricle, and because it contains all 3 layers of the cardiac wall, the acoustic properties are similar to the ventricular wall. A narrow neck and contractility are typical characteristics of congenital muscular ventricular diverticulum. Pulse-wave Doppler may reveal the biphasic waveform of the ejection flow in the presystolic and systolic phases at the ostium of the diverticular cavity.^{14,23} Contrast echocardiography can be used to evaluate intracardiac blood flow and myocardial perfusion, and to detect the muscular nature of the diverticular wall.^{13,23} Ventriculography is another reliable diagnostic method,²³ but its invasive character makes it less acceptable than echocardiography. Magnetic resonance imaging²⁴ and multiple slice computed tomography scanning may also allow accurate detection, but they are not as readily available as echocardiography.

The natural history of congenital cardiac diverticula is often determined by the severity of its associated abnormalities. Most patients will remain asymptomatic with no complications during long-term follow-up, suggesting that the clinical course may be benign.¹⁰ However, some fatal complications have been reported. Spontaneous rupture of a diverticulum has been found in some patients with sudden death.^{8,25} Our 4-year-old patient survived a rupture into the pericardium. Recurrent and refractory tachyarrhythmias, including ventricular tachycardia and fibrillation, may be another cause of sudden death. Systemic embolism, conges-

tive heart failure, and severe valvular regurgitation have also been reported. In our 30-year-old patient, severe valvular regurgitation was probably due to prolapse of an aortic valve leaflet caused by the subaortic diverticulum.

The treatment of congenital cardiac diverticula should depend on the clinical condition of the patient and any associated abnormalities. Surgical excision of a congenital cardiac diverticulum is usually performed in association with the correction of other cardiac abnormalities. Asymptomatic isolated congenital cardiac diverticula present a management dilemma. Some institutes advocate early surgical intervention for all diagnosed cardiac diverticula, even without clinical symptoms, to avoid potential risks such as rupture, systemic thromboembolism, arrhythmia, endocarditis, or sudden death.^{2,8,26} Other authors, however, recommend conservative therapy with close follow-up because of the benign prognosis in most cases, particularly in cases of muscular diverticula that may be less likely to rupture than fibrous diverticula.^{9,10} If there are refractory symptoms, significant complications, or a rapid increase in size during follow-up, elective surgery should be undertaken.

In our first case involving the 5-year-old boy, the fibrous subaortic diverticulum was complicated by rupture, pseudoaneurysm formation, and a hemopericardium. The blood was evacuated in the first surgical procedure, and the pseudoaneurysm was repaired in the second procedure, but it was only in the third surgical procedure that the diverticulum was recognized and resected. Although the sequential surgical procedures were uneventful and at the 6-month follow-up the boy was well, the process was difficult for both the boy and the surgeons. After this experience, surgical intervention was considered mandatory and proved successful for the 30-year-old man with a fibrous subaortic diverticulum complicated by endocarditis and both aortic and mitral regurgitation. Both cases 2 and 4 had severe aortic regurgitation requiring surgical intervention. At 1-year follow-up, all patients were well.

Conclusions

Congenital cardiac diverticulum is a rare anomaly. Its exact cause and prognosis remain unclear. TTE and transesophageal echocardiography are reliable tools for the diagnosis and follow-up. Multislice computed tomography and magnetic resonance imaging are also useful tools for the same purpose. Isolated congenital cardiac diverticula tend to involve a benign clinical process, but may be complicated by spontaneous rupture, systemic thromboembolism, arrhythmia, endocarditis, or sudden death. Surgical resection is performed when other associated congenital cardiac anomalies require surgical correction or complications require surgical therapy, but surgery is not mandatory for asymptomatic patients with isolated congenital cardiac diverticula.^{17,23,24}

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