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Original Article

Echocardiographic presentations of endocarditis, and risk factors for rupture of a sinus of Valsalva in childhood

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Abstract *Background:* In recent years, the diagnosis of infective endocarditis has been enhanced by the use of echocardiography. We sought, therefore, to review its effect on the management of endocarditis in children. *Methods*: We reviewed all the patients presenting to our institution for evaluation for infective endocarditis from May 1994 to January 2002. The patients were stratified according to whether or not they had congenitally malformed hearts. *Results:* Of the 90 referred patients identified, 46 (51%) had positive ultrasonic findings. Of these, we excluded 26 patients because of the presence of indwelling lines. The remaining 20 patients with features of endocarditis had a median age of 6.5 years, and a range from 0.14 to 8.5 years. There were 4 patients with normal hearts, and 16 with congenital cardiac malformations. We identified rupture of a sinus of Valsalva in four patients, with rupture into the left ventricle in two, and into the right ventricle and right atrium in one each. The mitral valve was involved in six patients, the aortic valve in another six, including all four with rupture of the sinus of Valsalva, both mitral and aortic valves in three, the pulmonary trunk in three patients, and the tricuspid valve and a Blalock-Taussig shunt in one patient each. Organisms isolated included Streptococcus mitis in 4 patients, Streptococcus pneumoniae in 2 patients, Streptococcus sanguis in 1, Staphylococcus aureus in 3, Staphylococcus epidermidis in 1, and Enteroccocus in 2. Cultures proved negative in 7 patients. Surgical intervention was needed in 12 patients, and one died (5%). Only the left-sided chambers were involved in those with normal hearts. Both patients infected with Streptococcus pneumoniae had rupture of a sinus of Valsalva. *Conclusion:* Involvement of the left-sided chambers is more likely in structurally normal hearts, and in cases with rupture of a sinus of Valsalva, in which case infection with Streptococcus pneumonia should be suspected.

Keywords: Bacterial infection; surgical intervention; streptococcus pneumoniae

Infective endocarditis is a life-threatening disease that is relatively rare in children, with a reported prevalence ranging from 1 in 1280 to 1 in 4500 hospital admissions.[1,2] Its definition was initially based on the classical criterions proposed by Van Reyn in 1881.[3] Secondary to the various changes in risk factors, epidemiology, and particularly the introduction of echocardiography, the criterions were expanded introduced in 1994, including echocardiographic diagnosis as one of the major criterions.[4] The purpose of our study was to review recent changes in echocardiographic presentations, and the effect of echocardiographic diagnosis on the management of infective endocarditis in children.

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Patients and methods

We performed a retrospective review of all patients presenting to the Texas Children's Hospital echocardiography laboratory for evaluation for infective endocarditis from May 1994 to January 2002. The clinical records were reviewed for evidence of congenital cardiac disease, timing and presentation of infective endocarditis, prior surgical or catheter-based intervention, and to extract the positive echocardiographic findings.

The microbiological records were reviewed to determine the organism isolated. Patients were divided into two groups, those with and without a congenital cardiac malformation.

Results

Among the 90 patients identified from the echocardiography database who underwent praecordial echocardiography for evaluation of infective endocarditis, positive echocardiographic findings were found in 46 (51%). Of these, 26 had indwelling central lines and were excluded, leaving 20 with echocardiographic findings consistent with infective endocarditis. In these twenty patients, the median age was 6.5 years, with a range from 0.14 to 8.5 years.

The hearts were structurally normal in 4 patients, with the other 16 having associated congenital cardiac malformations. The median age was comparable in the two groups, being 5.8 years, with a range from 2.3 to 7.8 years, in those with normal hearts, and 6.5 years, with a range from 0.14 to 8.5 years in those with congenital cardiac malformations. Of the four patients with normal hearts, only one had an identifiable origin of endocarditis, a septic hip with positive blood cultures for Staphylococcus aureus. Only one other of these patients had a patent oval foramen. The three patients with no obvious origin of infection also had no history of dental extraction or body piercings. The mitral valve was involved in all four patients and the aortic valve in one patient, the latter having rupture of a Valsalva.

In the 16 patients with malformed hearts, 6 had a ventricular septal defect (37%), valvar aortic stenosis was present in 3 patients (18%), valvar pulmonary stenosis in another 3 patients (18%), and one patient each (6%) had atrioventricular septal defect with common atrioventricular valve, double inlet left ventricle and transposition of the great arteries (concordant atrioventricular with discordant ventriculo-arterial connections). In the final patient, there was residual shunting through an arterial duct subsequent to attempted closure with a 0.52-inch coil.

Of the patients with congenital cardiac lesions, a procedure had occurred within one month of diagnosis in four. Surgical repair of a perimembranous ventricular septal defect, aortic valve stenosis, and transposition had been performed in one each, with the fourth patient having had attempted transcatheter occlusion of the patent arterial duct. A pericardial patch had been used for closure of the ventricular septal defect, a homograft was used for repair of the aorta, and no foreign material had been in the repair of the child with transposition.

Endocarditis was diagnosed between three weeks and ten weeks after intervention in all patients. No patient who developed endocarditis in a congenitally malformed heart had a history of preceding body piercing, although one patient with aortic valvar stenosis had undergone a dental extraction four weeks prior to the development of endocarditis. This patient had received appropriate antibiotic therapy at the time of dental extraction.

Echocardiographic findings

Precordial transthoracic echocardiography was performed in all patients, and transesophageal recordings had been made in 12 patients who underwent surgical resection of vegetations or valvar repair or replacement. Rupture of an aortic sinus of Valsalva occurred in four patients. Of these four, one patient had undergone a Rastelli procedure for repair of double outlet right ventricle, and was immunocompromised secondary to prior renal transplantation. A ventricular septal defect had been closed in the second patient, and the aortic valve replaced with a homograft in the third. The heart was structurally normal in the fourth patient. In two of these cases, the sinus had ruptured into the left ventricle. The others had ruptured into the right atrium and the right ventricle, respectively (Figs 1 and 2).

The mitral valve was involved in six patients, and the aortic valve in six, including the four patients with rupture of a sinus of Valsalva. Both mitral and aortic valves were involved in three patients, the tricuspid valve in one patient, the pulmonary trunk in three patients (Fig. 3), and a Blalock-Taussig shunt in one patient.

Suspicion of infective endocarditis was raised in three patients (15%) by echocardiographic examination prior to clinical suspicion or the confirmation of positive blood cultures. All these patients had undergone previous surgery, specifically for closure of a ventricular septal defect, insertion of an aortic homograft, or construction of a Blalock-Taussig shunt.

Microbiology

The organisms isolated by blood culture included Streptococcus mitis in 4 patients, Staphylococcus aureus in three patients, Streptococcus pneumoniae in two patients, Enterococcus in two patients, Staphylococcus epidermidis in one, and Streptococcus sanguis in one patient. In the remaining seven patients, blood cultures were negative (Fig. 4).

Course and management

Surgical intervention was needed in 12 patients, including surgical debridement of the vegetations. When valvar dysfunction or regurgitation was severe,

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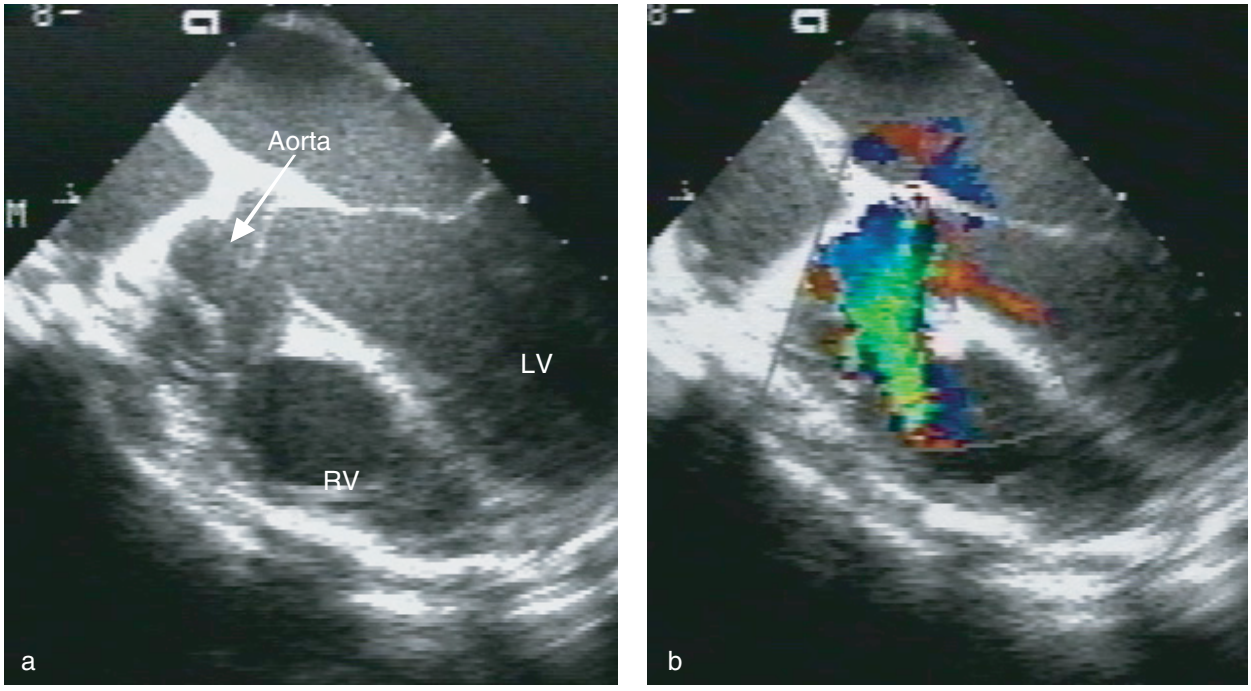

Figure 1.
The transesophageal echocardiogram demonstrates rupture of an aortic sinus of Valsalva into the right ventricle.

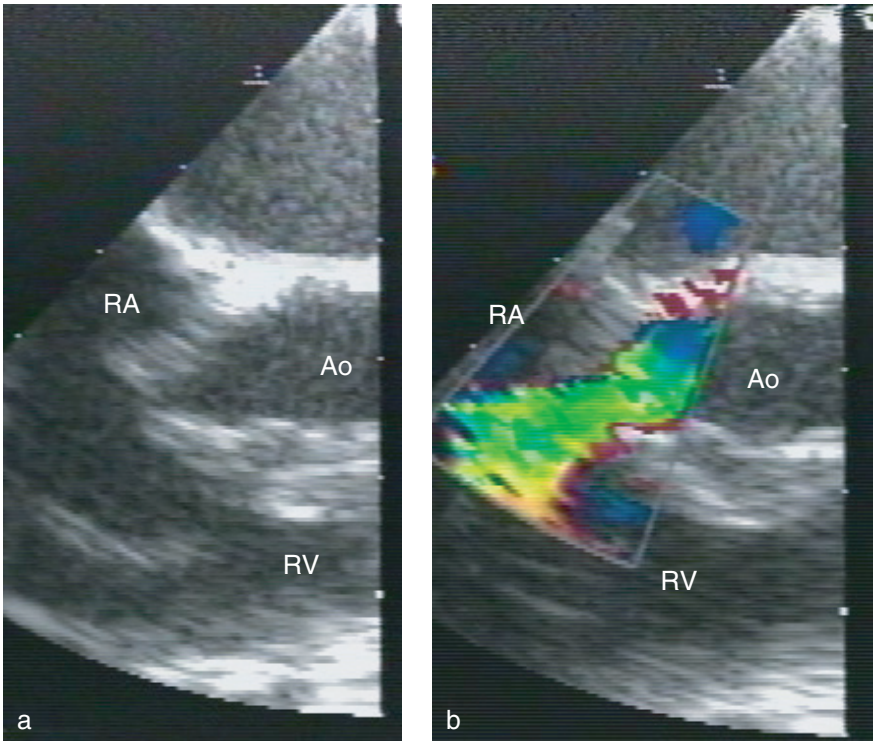

Figure 2.
This transesophageal echocardiogram demonstrates rupture of an aortic sinus of Valsalva into the right atrium.

the valve involved was replaced. This was the aortic valve in five patients, all having a homograft inserted, including all four children with rupture of a sinus of Valsalva. The mitral valve was repaired or replaced in another five patients, with one patient requiring replacement of both the aortic and mitral valves. The final patient had resection of vegetations involving the right ventricular outflow tract and the pulmonary trunk. Surgery was required at a median of five weeks from diagnosis, with a range from two days in a child with severe aortic regurgitation secondary to rupture of an aortic sinus of Valsalva, to

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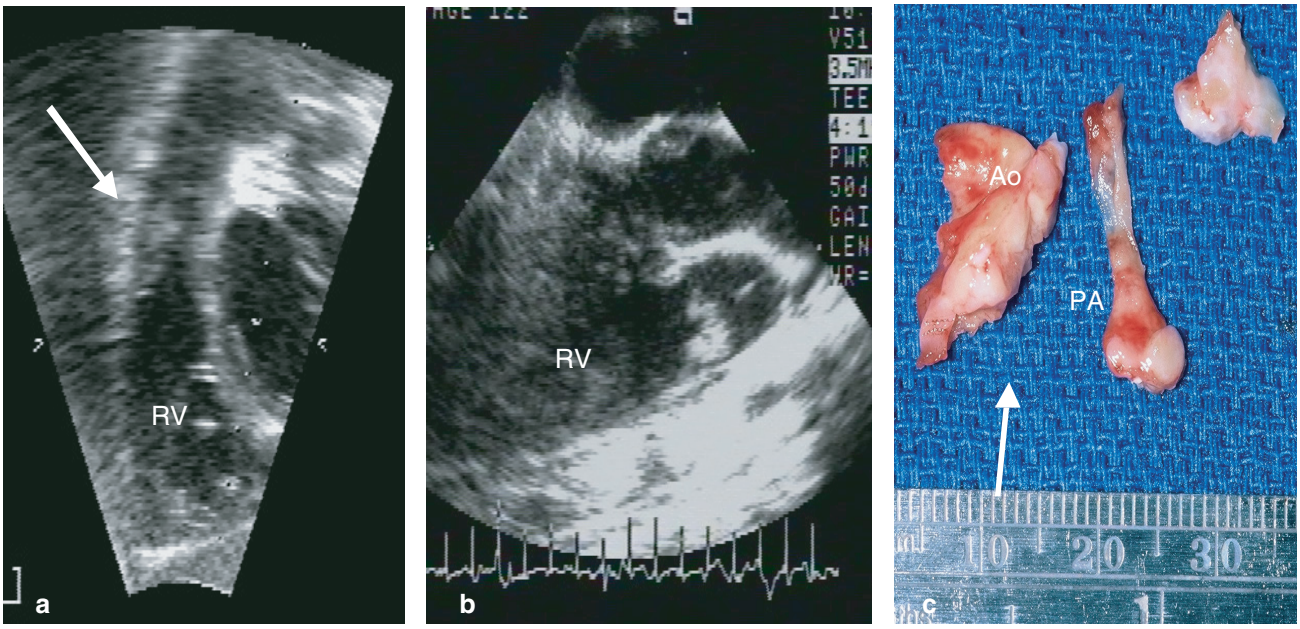

Figure 3.
The transesophageal echocardiogram demonstrates vegetations on the pulmonary valve and in the right ventricular outflow tract (a,b), with panel c showing the gross pathology of the vegetations extracted at surgical debridement.

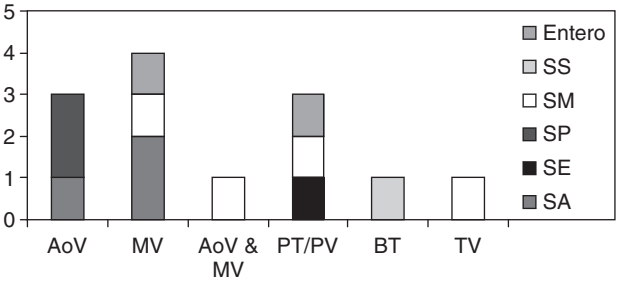

Figure 4.
The bar chart demonstrates the distribution of organism and site of vegetations. The x-axis represents the number of patients. AoV: aortic valve; BT: Blalock-Taussig shunt; MV: mitral valve; PT: pulmonary trunk; PV: pulmonary valve; TV: tricuspid valve; Entero: Enterococcus; SS: Streptococcus sanguis; SM: Streptococcus mitis; SP: Streptococcus pneumoniae; SE: Staphylococcus epidermidis; SA: Staphylococcus aureus.

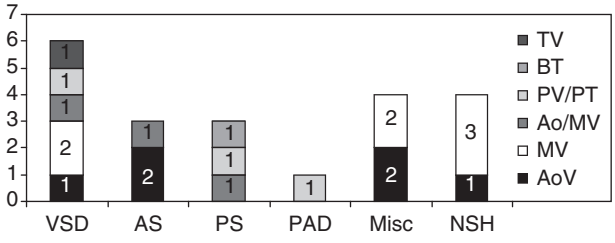

Figure 5.
The bar chart demonstrates the distribution of the site of vegetations and the associated cardiac malformations. AS: aortic valvar stenosis; Misc: miscellaneous; NSH: no surgical history; PS: pulmonary stenosis; PAD: patent arterial duct; VSD: ventricular septal defect.

three months in a patient with progressive development of severe mitral regurgitation.

All patients underwent standard intravenous antimicrobial therapy for six weeks based upon the sensitivities of the isolated organism. Death occurred in one patient (5%), who had undergone repair of a perimembranous ventricular septal defect and developed severe aortic regurgitation due to involvement of the valve in the infective process, with subsequent left ventricular failure and failure of multiple organs.

There were no cases of recrudescent endocarditis, neither after surgical replacement nor medical management of the endocarditis, including the patients requiring surgery early in the course of their disease.

Correlation of organism with valvar involvement

The distribution of the site of the vegetation and the associated cardiac malformations are demonstrated in Figure 4, with the various organisms and the site of vegetation correlated in Figure 5. Those patients with structurally normal hearts had involvement only of the left-sided valves, with the mitral valve involved in three patients and the aortic valve involved

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in the other. Both patients infected by Streptococcus pneumoniae developed rupture of an aortic sinus of Valsalva.

Discussion

Echocardiographic findings were added as a major criterion for diagnosis of infective endocarditis in 1994, despite contemporaneous reports of a relatively low yield following echocardiographic investigation in the absence of high clinical suspicion and positive blood cultures.[4,5] The yield for identifying vegetations in a patient with positive blood cultures has been reported to be higher if the organism isolated was Staphylococcus aureus, a fungal organism, if the patient was premature, immunocompromised or had an indwelling catheter.[6] In our study, half of the patients had positive echocardiographic findings, demonstrating that improvements in resolution of equipment over recent years has led to a change in the incidence and yield of echocardiographic investigation in the diagnosis of infective endocarditis. This is further supported in our study by the establishment of the diagnosis in patients with otherwise normal hearts, and in three patients prior to evidence of bacteremia.

Echocardiographic visualization of vegetations has been shown to identify a sub-group of patients at high risk.[7,8] Mortality from infective endocarditis was reduced from 30% in 1983[9] to 2.6% in 1992,[8] a trend that is supported by our own experience. The presence of a predisposing cardiac anomaly is considered one of the primary elements for diagnosis of infective endocarditis in both the original criterions of Von Reyn[3] and the modifications offered by the group from Duke University.[4] In our children, however, four patients (20%) had no documented cardiac disease, similar to the proportion of 17% reported by Van Hare and colleagues.[1] The distribution of valvar involvement mirrors that reported by Rohmann and colleagues,[8] who found vegetations on the aortic valve in three-fifths, and on the mitral valve in two-fifths of their cohort.

In our children, those with structurally normal hearts only had involvement of the left-sided valves, with a particular predisposition to involvement of the mitral valve. This is difficult to rationalize, as only one of these patients had a patent oval foramen. Streptococcus pneumoniae was cultured only in these patients with otherwise normal hearts but endocarditis of the aortic valve, which differs from previous studies showing Staphylococcus aureus to be the predominant organism infecting the aortic valve.[8] Rupture of a sinus of Valsalva in our children was more likely to be associated with Streptococcus pneumoniae bacteremia. In those patients with ruptured sinuses of Valsalva, two had Streptoccocus pneumoniae isolated by blood culture, one had Staphylococcus aureus, and the culture was negative in the other. Both children with positive cultures for Streptococcus pneumoniae experienced rupture of a sinus of Valsalva. Previously, Staphylococcus aureus has been considered the predominant organism associated with this kind of rupture.[10]

Limitations

The major limitation of our study is its retrospective nature, which has well recognised inherent limitations. The relatively small number of patients made statistical analysis difficult, but due to the relatively rare nature of this disease process nowadays it is difficult to study a large group of patients.

Conclusion

Echocardiographic diagnosis of infective endocarditis was higher in children than previously reported, which reflects improvements in echocardiographic resolution in recent years. Involvement of the left-sided valves was the typical pattern in those patients with structurally normal hearts. Patients with Streptococcus pneumoniae bacteremia had a higher risk of rupture of an aortic sinus of Valsalva.

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