



A case report on endarteritis in a child with coarctation of aorta

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Abstract

Coarctation of aorta (CoA), complicated by endarteritis in children is very rare. Here we present a case of endarteritis in an unoperated CoA in a four year old boy. CoA had been diagnosed in the referring hospital, yet the diagnosis of endocarditis distal to CoA, was made in the tertiary center using modified transthoracic echo windows or focused views. After six weeks of intravenous antibiotic treatment, a coarctectomy and end-to-end anastomosis was performed and he recovered clinically well. This case report concludes that echocardiography remains as the standard diagnostic method for identifying intracardiac manifestations of infective endocarditis/endarteritis. Last but foremost, it delineates the importance of modified transthoracic echo windows or focused views in identifying the unusual position of endocarditis.

KEYWORDS

coarctation of aorta, congenital heart disease, endarteritis, infective endocarditis, *Streptococcus sanguinis*

1 | BACKGROUND

It has been proposed that two conditions should be met for the diagnosis of endocarditis: (a) the presence of damaged or traumatized endothelium, (b) entry of bacteria into the bloodstream.¹ The most commonly endocarditis in congenital heart disease is seen within and around heart valves, or structures adherent to prosthetic materials in postoperative conditions. In coarctation of aorta (CoA), blood flows through a narrowing in the aorta at high velocity, resulting in a lower pressure "sink" in the area distal to the stenosis. Bacteria may attach to the aortic wall in this low pressure region, especially when there is concurrent endothelial injury. In CoA, endothelial injury is likely to be precipitated by shear stress force.^{2,3} The bacterial pathogens that cause aortic endarteritis are the same pathogens which are known to cause valvular endocarditis including viridians group *Streptococci*, coagulase-negative *Staphylococci*, HACEK species:

Haemophilus species, *Aggregatibacter* species, *Cardiobacterium hominis*, *Eikenella corrodens*, and *Kingella* species, and *Staphylococcus aureus*.⁴ Aortic endarteritis is very rare in children.

2 | CASE PRESENTATION

A 4-year-old, previously healthy boy was diagnosed with Henoch-Schonlein purpura after an upper airway infection. A week later, he was admitted to a secondary hospital with fever and a painful right knee. He had dental enamel disorders. In addition, on auscultation a systolic murmur was heard. Blood cultures showed a positive growth of *Streptococcus sanguinis*. An echocardiography revealed a CoA without vegetations. Ultrasound and Magnetic Resonance Imaging of the right knee were normal. C-reactive protein (CRP) at initial presentation was 40 mg/L. He was diagnosed with a low-grade bacteremia and reactive arthritis

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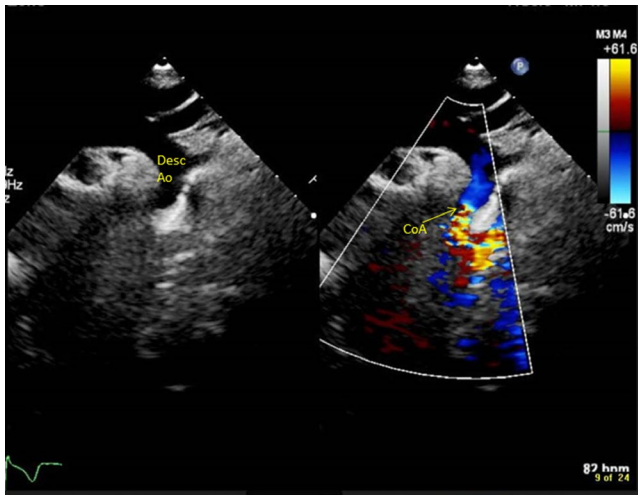


FIGURE 1 Suprasternal view of aortic arch showing coarctation of aorta (CoA). Yellow arrow indicates CoA. Desc Ao = descendens aorta

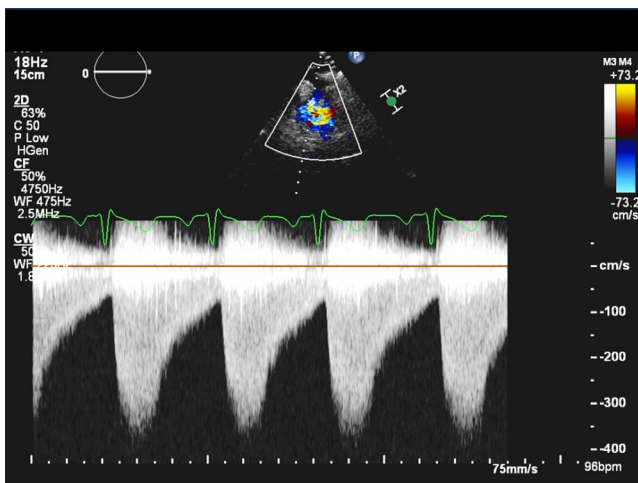


FIGURE 2 Doppler of coarctation of aorta. Continuous wave doppler in distal aorta descendens with continuous antegrade flow throughout diastole

and was treated with Penicillin. Despite treatment with antibiotics, his fever did not disappear and he developed abdominal pain. Blood cultures remained positive for *Streptococcus Sanguinis*, and CRP increased to 57 mg/L. Erythrocyte sedimentation rate (ESR) was 58 mm/h. Kidney function and urine sedimentation were normal. The abdominal ultrasound showed splenomegaly. Due to persisting bacteremia, abdominal pain, splenomegaly, and recurrent fever, he was referred to our tertiary heart center, approximately 6 weeks after the initial symptoms.

2.1 | Physical examination at admission

Physical examination revealed weak and delayed pulses in the lower extremities, a difference in blood pressure between upper and lower extremities more than 30 mm Hg, systolic ejection murmur grade 2/6, maximum at 2nd right intercostal space and radiating to back. Normal respiratory sounds, no hepatomegaly, and no tenderness in

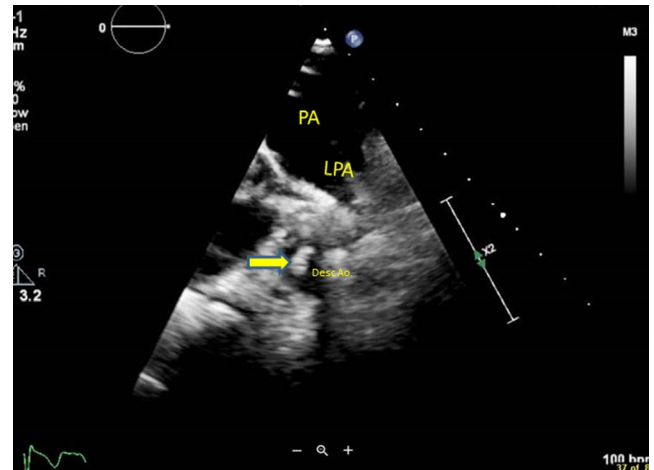


FIGURE 3 Modified ductal view in transthoracic echocardiography (TTE). Modified ductal parasternal echo window in TTE: Yellow bold arrow indicates vegetation in distal aorta descendens. Desc Ao = descendens aorta; LPA = left pulmonary artery; PA = pulmonary artery

abdomen were observed. He had no lymphadenopathy, but he was limping in his right leg.

Blood tests showed an increased levels of Immunoglobulin G (22.3 G/L), CRP (124 mg/L), and ESR (116 mm/h). The levels of hemoglobin were 5.7 mmol/L and leukocyte count $11.8 \times 10^9/L$. Three consecutive blood cultures were positive for *Streptococcus sanguinis*.

Transthoracic echocardiography (TTE): CoA of the distal aorta descendens. An echo dense structure was seen distal to the coarctation using nonstandard echo views, and a dubious echo dense structure was seen adjacent to the aortic valve (Figures 1-3). Movie S1, Movie S2 and Movie S3

Transesophageal echocardiography (TEE): a highly mobile vegetation was seen in the distal aortic arch. The aortic valve cusps were tricommisural, asymmetric with mild regurgitation and free of vegetation. (Figures 4-6, Movie S4, Movie S5 and Movie S6).

A Proton Emission Computed Tomography scan was performed to rule out any metastatic involvement of endocarditis in the body. There was no suspicion of active endocarditis in the heart and large vessels, no active inflammation in the knee or dental structures.

The child received 6 weeks of intravenous penicillin. Balloon dilatation of the CoA segment was not an option of treatment in this child, because of the risk of dislodging the remaining vegetation in the aorta descendens. After 6 weeks, he was operated through a left lateral thoracotomy. The coarctation segment was excised, a vegetation was removed, and an end to end anastomosis of aorta descendens was performed, together with the ligation of ductus segment and ligation of one of the collaterals from aorta descendens. He recovered well in postoperative period.

3 | DISCUSSION

In reviewing the literatures, we found 11 case reports including adults with CoA complicated with endarteritis. Out of these 11 case reports,

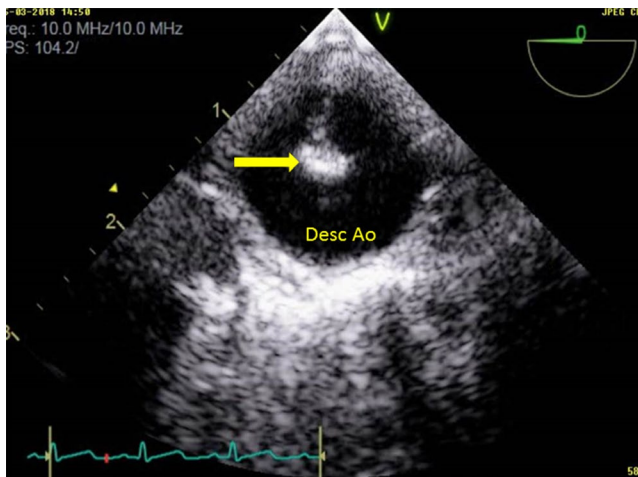


FIGURE 4 Transesophageal echocardiography - Aorta descendens. Yellow arrow indicates vegetation in distal aorta descendens at an angle of 0°. Desc Ao = descendens aorta

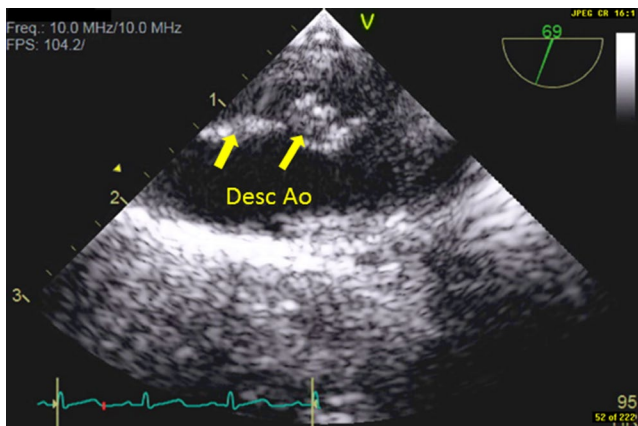


FIGURE 5 Transesophageal echocardiography (TEE)-Vegetation seen in distal of aorta descendens. Yellow arrows indicate highly mobile vegetation in distal aorta descendens, while angling the TEE probe. Desc Ao = descendens aorta

4 were children diagnosed with CoA and unoperated at the time of presentation. Thus, endarteritis in CoA is very rare entity in pediatric group. In 1946, Leininger et al⁵ reported a 10-year-old boy was diagnosed with CoA and superimposed bacterial endocarditis, 3 other patients/case reports had endarteritis and mycotic aneurysm.⁶⁻⁸ One patient had pseudo aneurysm and endarteritis after resection of coarctation of aorta.⁹ In a 60 year single institution retrospective review from the Mayo clinic, *Viridans streptococci* and *Staphylococcus aureus* were the most common pathogens causing endocarditis.¹⁰ The high velocity from abnormal blood jet stream, the flow from high pressure to a low pressure chamber, and the presence of narrow orifice between two chambers or blood vessels capable of creating a pressure gradient, facilitates a hemodynamic situation which causes endothelial damage of the artery.¹ Clinical presentation of infective endocarditis can be acute, rapidly progressive infection, but also as subacute or chronic with low-grade fever and nonspecific symptoms that may mislead or confuse initial assessment. A high index of suspicion and low

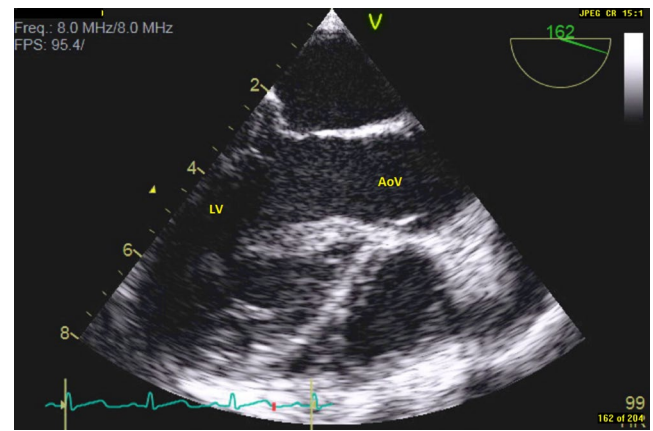


FIGURE 6 Aortic valve free of vegetation in transesophageal echocardiography. Ao = aorta, LV = left ventricle

threshold for investigations is needed in high-risk groups such as patients with congenital heart disease (CHD) or prosthetic valves to rule out infective endocarditis or avoid delay in diagnosis.^{11,12} Reifenstein et al¹³ reported in a review that 70% out of 104 autopsied cases of CoA and bacterial endocarditis had bicuspid valves, interestingly in our case report, aortic valve is tricuspid and asymmetrical. In all other pediatric case reports described earlier, endarteritis was diagnosed by transesophageal echocardiography or by CT or MRI, whereas this case report is the first where endarteritis was diagnosed with transthoracic echocardiography.

4 | CONCLUSION

Endarteritis complicating CoA is rare and should be suspected in pediatric patients with relevant clinical signs and symptoms. Echocardiography remains the standard diagnostic method for identifying intracardiac manifestations of infective endocarditis/endarteritis and plays a key role in the diagnosis, management, and monitoring of these patients. Performing modified echo windows or focused views will definitely help in imaging the unusual location of endocarditis.

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CONFLICT OF INTEREST

The authors have no conflicts of interest.

ETHICAL APPROVAL

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

This article does not contain any studies with animal participants performed by any of the authors.

INFORMED CONSENT

Parents or their legal representatives provided written informed consent in accordance with guidelines.

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SUPPORTING INFORMATION

Additional supporting information may be found online in the Supporting Information section at the end of the article.

Movie S1. Suprasternal view of aortic arch showing coarctation of aorta (CoA). Yellow arrow indicates CoA. Desc Ao = descendens aorta.

Movie S2. Modified ductal view in transthoracic echocardiography. Modified ductal parasternal echo window: yellow bold arrow indicates vegetation in distal aorta descendens. Desc Ao = descendens aorta; LPA = left pulmonary artery; PA = pulmonary artery.

Movie S3. Modified ductal view in color. Yellow bold arrow indicates vegetation in distal aorta descendens. Desc Ao = descendens aorta; LPA = left pulmonary artery; PA = pulmonary artery; RPA = right pulmonary artery.

Movie S4. Transesophageal echocardiography (TEE) - Aorta descendens. Yellow bold arrow indicates vegetation in distal aorta descendens at an angle of 0°. Desc Ao = descendens aorta.

Movie S5. Transesophageal echocardiography (TEE)- Vegetation seen in distal of aorta descendens. Yellow arrows indicates highly mobile vegetation in distal aorta descendens, while angling the TEE probe. Desc Ao = descendens aorta.

Movie S6. Aortic valve free of vegetation in transesophageal echocardiography. Ao = aorta; LV = left ventricle.

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