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GUEST EDITOR'S PAGE

Endocarditis in ACHD, Be Aware, PREVENT, Diagnose Early and Treat

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Patients with congenital heart disease (CHD) represent an ever-growing patient cohort owing to advancements in medical and surgical treatment. Depending on the underlying level of CHD complexity, this group of patients can have multiple needs over their lifetime for which long-term follow-up is warranted.[1] As adult congenital cardiologists, it is vital that we do not just monitor and treat any residual lesions or symptoms but aim to empower our patients and strive toward prevention whenever possible.[2]

Infective endocarditis (IE) is a leading cause of morbidity and mortality among patients with CHD. An estimated incidence of 1.33 cases per 1,000 person-years was reported in a study by Kuijpers et al[3] looking at 14,224 patients with CHD, a value 27 to 44 times greater than the general adult population. This increased predisposition to IE is owed in part to the prosthetic materials used for repair or palliative procedures and/or residual defects such as small ventricular septal defects or valvular dysfunction.[3-5] Valve-containing prosthetics was shown to be a significant independent risk factor for IE both in the short- and long-term, corroborating the European Society of Cardiology/American Heart Association guidance on antibiotic prophylaxis. This is in contrast to nonvalve-containing prosthetics, in which risk is greater in the first 6 months' post-implantation where prophylaxis is again recommended.[3,6] In the United Kingdom, the National Institute for Health Care and Excellence published guidance in 2008 advising against any routine antibiotic prophylaxis.[7] A study by Dayer et al[8] in the *Lancet* in 2015 showed there was a significant increase in IE cases after withdrawal of antibiotic prophylaxis, although a causal relationship could not be established.

IE accounts for up to 4% of admissions to tertiary adult congenital heart disease (ACHD) services, with a reported mortality of 6.9% to 8%.[9-12] Recurrent endocarditis was also shown to be common in patients with ACHD; for example, in a recent study of 164 patients from our center, 23% had had previous episodes of IE. Predisposing factors were only identified in 26.2%, with the most common being dental procedures (27.9%) followed by a skin infection (23.3%). Other less common potentially predisposing events were recent surgical or catheter procedures, gynecologic interventions, and upper respiratory tract, gastrointestinal, or urinary tract infections. Thirty-seven percent of patients required a surgical intervention on their index admission in our experience.[10] Although patients with ACHD differ in terms of anatomical complexity, the majority remain at risk of IE, making prevention an even more important strategy.

A study by Bauer et al[13] in 2017 examined 1,211 patients with ACHD from the German National Register for Congenital Heart Disease evaluating levels of knowledge and understanding on the topic of IE. Of those patients, 74.5% (902) reported they were aware of what IE was but only 76.5% (690) selected the correct definition on the multiple-choice survey. Knowledge about IE antibiotic prophylaxis was exhibited by just 55.4% (672) of the patients analyzed. This study highlights the knowledge gaps on this life-threatening condition that can affect patients with ACHD and the pressing need to address this gap.

Along the same lines, a recent retrospective study by Brida et al[14] from the study group on ACHD in Central and South Eastern Europe looked at 295 patients with ACHD and evaluated diagnosis and outcomes. This study found a median time of 25 days

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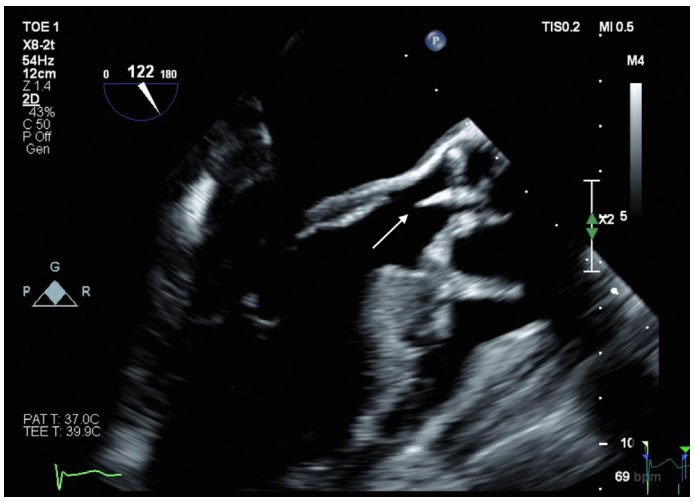
FIGURE 1 Transesophageal Echocardiogram



Mid esophageal 3-chamber view showing aortic valve vegetation **(white arrow)**.

between onset of symptoms and diagnosis, with an alarming 68.8% of patients receiving empirical antibiotics before essential investigations (eg, blood cultures) toward establishing the diagnosis and identifying causative organisms. Although survival in this cohort of patients was similar to that of other European countries, the need to educate patients with ACHD and colleagues not trained in cardiology/ACHD was clearly evident regarding early diagnosis and timely treatment of IE.

We highlight all these important issues around patient understanding, prevention, and treatment of IE with a recent case from our center. This case involved a 56-year-old man with a background history of bicuspid aortic valve, subaortic stenosis, and 3 previous sternotomies, including 2 subaortic resections and aortic valve repair with aortic root enlargement. Other medical history included chronic kidney disease (glomerular filtration rate of 37 mL/min), hypercholesterolemia, and eczema. The patient presented to our hospital for routine outpatient review with a 6-week history of night sweats and a weight loss of 20 kg in the preceding 2 months. He also reported flu-like symptoms with cough, sickness, and nausea in the previous few months, for which he was seen by his general practitioner and was given a short 5-day course of oral antibiotics. IE was not considered during that time, and neither blood tests nor blood cultures were conducted. Antibiotics improved the symptoms temporarily before recurring; the patient went to his local emergency department, where similarly no blood cultures were drawn, and the patient was given a further course of antibiotics for a chest infection. The patient did not recognize any of these as potential features of subacute IE nor did he contact our team until his planned review later in the year. He maintained good dental hygiene throughout and saw a dentist earlier that year.

Initial blood cultures in our hospital grew *Streptococcus oralis*, and bedside echocardiography revealed mobile vegetations on the noncoronary cusp of the aortic valve and on the anterior leaflet of the mitral valve. There was severe aortic regurgitation with moderate to severe mitral regurgitation. Transesophageal echocardiography confirmed these findings (Figure 1). Magnetic resonance imaging of the patient's brain showed a possible small septic embolus with no evidence of surrounding edema or ischemia; abdominal imaging showed no evidence of further septic emboli in the systemic bed. The patient received a 2-valve replacement operation and a fourth sternotomy after an initial course of intravenous antibiotics with a good and uncomplicated postoperative course.

It is clear more work needs to be done in terms of prevention of IE in general and in the CHD-vulnerable group. The usual source of bacteria entry is the mouth (streptococci) and the skin (staphylococci). Optimal mouth and skin hygiene, healthy diet, not smoking, brushing and flossing, not biting nails, avoiding tattoos, and prompt treatment of paronychia or other skin infections are bound to minimize the risk of IE. A low threshold for symptoms and signs of IE by the patients themselves and resistance to over-the-counter antibiotic prescription before blood cultures are drawn are likely to improve outcomes (Figure 2).

Last but not least, being slim and athletic, our new aspirational mantra, can serve as a boost to our immune system in general, conveying protection or better response to IE, coronavirus, or any other infection. We must do better in the future; if not, it would be at our patient's peril.

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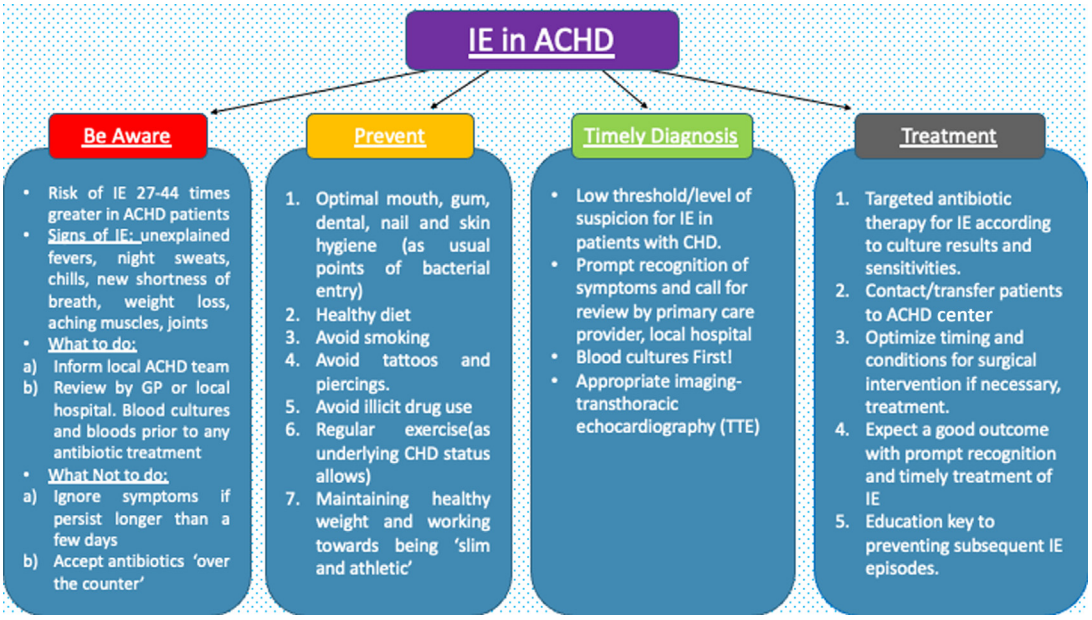
FIGURE 2 **IE in ACHD**



Take-home messages to improve awareness, prevention, timely diagnosis, and treatment of patients with adult congenital heart disease (ACHD). IE = infective endocarditis.

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