



## Diagnostics

# The pediatric electrocardiogram Part III: Congenital heart disease and other cardiac syndromes

Matthew O'Connor MD<sup>a</sup>, Nancy McDaniel MD<sup>b</sup>, William J. Brady MD<sup>c,\*</sup>

<sup>a</sup>Department of Pediatrics, Childrens' Medical Center, University of Virginia Health System, Charlottesville, VA

<sup>b</sup>Cardiology Division, Department of Pediatrics, Childrens' Medical Center, University of Virginia Health System, Charlottesville, VA

<sup>c</sup>Department of Emergency Medicine, University of Virginia Health System, Charlottesville, VA

Received 27 July 2007; accepted 2 August 2007

**Abstract** Approximately 1% of newborns are affected by congenital heart disease (CHD), and although many lesions of CHD have trivial hemodynamic and clinical implications, some clinically significant lesions are asymptomatic in the immediate newborn period and may present after discharge from the well baby nursery. Because of this, CHD should be considered in the differential diagnosis of any ill-appearing newborn, regardless of the presence of cyanosis. In addition, the number of children, adolescents, and adults with surgically repaired or palliated CHD continues to grow within the United States and other developed countries. It is in this population that arrhythmias are particularly prone to develop, and knowledge of the common arrhythmias associated with CHD is mandatory for the acute care provider. © 2008 Elsevier Inc. All rights reserved.

## 1. Introduction

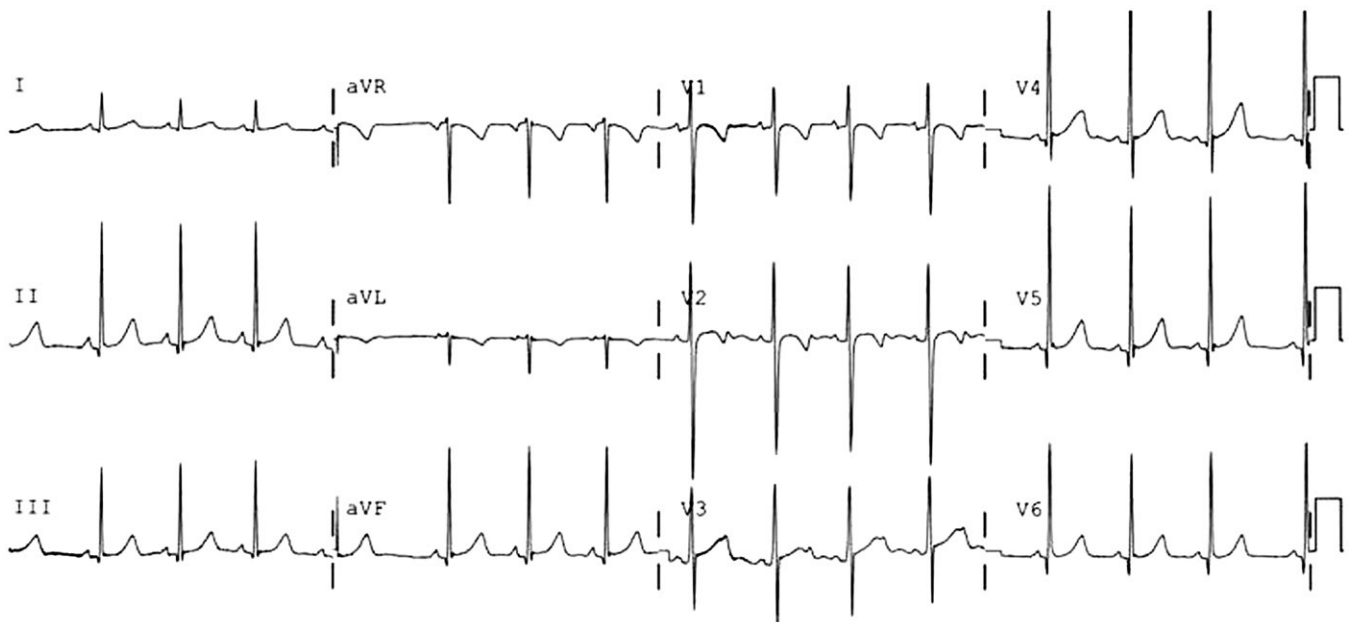
Approximately 1% of newborns are affected by congenital heart disease (CHD), and although many lesions of CHD have trivial hemodynamic and clinical implications, some clinically significant lesions are asymptomatic in the immediate newborn period and may present after discharge from the well baby nursery. Because of this, CHD should be considered in the differential diagnosis of any ill-appearing newborn, regardless of the presence of cyanosis. In addition, the number of children, adolescents, and adults with surgically repaired or palliated CHD continues to grow within the United States and other developed countries. In the United States, as of 2001, it has been estimated that nearly 1 million adults live with CHD

[1]. It is in this population that arrhythmias are particularly prone to develop, and knowledge of the common arrhythmias associated with CHD is mandatory for the acute care provider.

In infants and children with suspected CHD, the electrocardiogram (ECG), in combination with other invasive and noninvasive studies, has largely been supplanted by echocardiography as the modality of choice with regard to diagnosis. However, the ECG remains an important screening tool in the evaluation of infants and children with clinically suspected CHD. In addition, the ECG is frequently an adjunct to diagnosis in other forms of pediatric heart disease. In this final article of the series, common ECG abnormalities will be discussed with regard to 3 groups of patients: (1) infants and children with undiagnosed CHD, (2) infants and children with myocardial hypertrophy or ischemia, and (3) children and young adults with repaired or surgically palliated structural CHD.

\* Corresponding author.

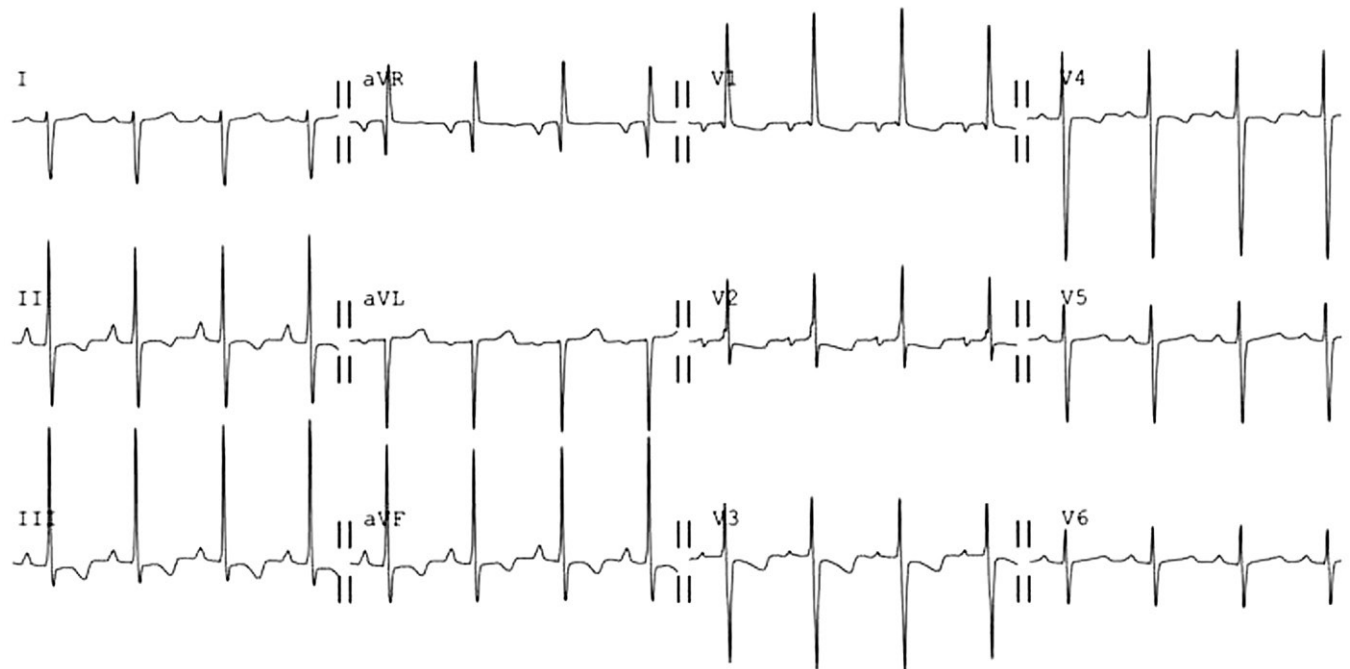
E-mail address: [wb4z@virginia.edu](mailto:wb4z@virginia.edu) (W.J. Brady).



**Fig. 1** LVH in a 3-year-old boy with unrepaired coarctation of the aorta. Note the deep S waves in V1 and V2 with tall R waves in V4 and V5.

The ECG is an insensitive and nonspecific screening method in the diagnosis of CHD, unless an arrhythmia is present [2]. A normal ECG does not rule out hemodynamically significant CHD. However, because it is noninvasive and inexpensive, it is frequently used in the initial evaluation of infants or children who present to the emergency department with symptoms suggestive of heart disease. The careful observer can infer much from a cursory

evaluation of the ECG. For example, the finding of left ventricular hypertrophy (LVH; Fig. 1), which should always be unusual in newborns, suggests the presence of left-sided obstructive lesions such as aortic stenosis or coarctation of the aorta, although in the presence of a VSD, right ventricular hypertrophy (RVH) may also exist (Fig. 2). In older children with characteristic findings on auscultation, LVH may be an indicator of hypertrophic obstructive cardiomyopathy. The



**Fig. 2** RVH in a 10-year-old boy with primary pulmonary hypertension. Note the tall R waves in V1 and V2 and the deep S waves in V5 and V6. Right axis deviation is also present.



**Fig. 3** Left atrial enlargement in a 9-month-old female infant with combined mitral and tricuspid insufficiency. Note the deep, broad p waves in lead V1 and tall, notched p waves in lead II.

diagnosis of RVH via ECG should be made cautiously in newborns, but after 6 months of age, right-sided obstructive lesions such as tetralogy of Fallot or abnormalities of the pulmonary vasculature (ie, pulmonary hypertension) should be considered in the differential diagnosis. It is important to note that infants with definitive RVH may have left-sided obstructive lesions such as aortic stenosis or coarctation of the aorta secondary to associated pulmonary hypertension.

Further analysis of p wave morphology may lend insights into the presence of CHD. The concepts of left and right atrial abnormality are important in gleanng information from the ECG. Left atrial abnormality simply implies left atrial enlargement, and as discussed in article 1 of this series, left atrial hypertrophy is manifested on the ECG by a deep (5 mm) biphasic p wave in V1 or a broadened p wave in lead II (Fig. 3). Left atrial abnormality may be seen in any condition causing left atrial volume overload; representative lesions include mitral stenosis or insufficiency, atrioventricular canal defects, and left ventricular failure from diverse causes. Right atrial abnormality similarly represents right atrial enlargement; it is defined as a p wave voltage of greater than 2.5 mm or duration greater than 100 milliseconds (Fig. 4). Right atrial abnormality may be seen in any condition with large left-to-right shunts or in right ventricular (RV) failure, as in atrioventricular canal defects, tricuspid atresia, atrial septal defects, or Ebstein abnormality of the tricuspid valve [2].

Abnormalities of axis are rarely specific for a certain condition. However, one specific instance of axis deviation should be mentioned because its presence classically indicates the presence of an atrioventricular canal defect. The so-called superior axis deviation, in which the axis is deviated to the extreme right ( $-90^{\circ}$  to  $180^{\circ}$  or  $+180^{\circ}$  to  $+270^{\circ}$ ), is frequently seen in patients with atrioventricular

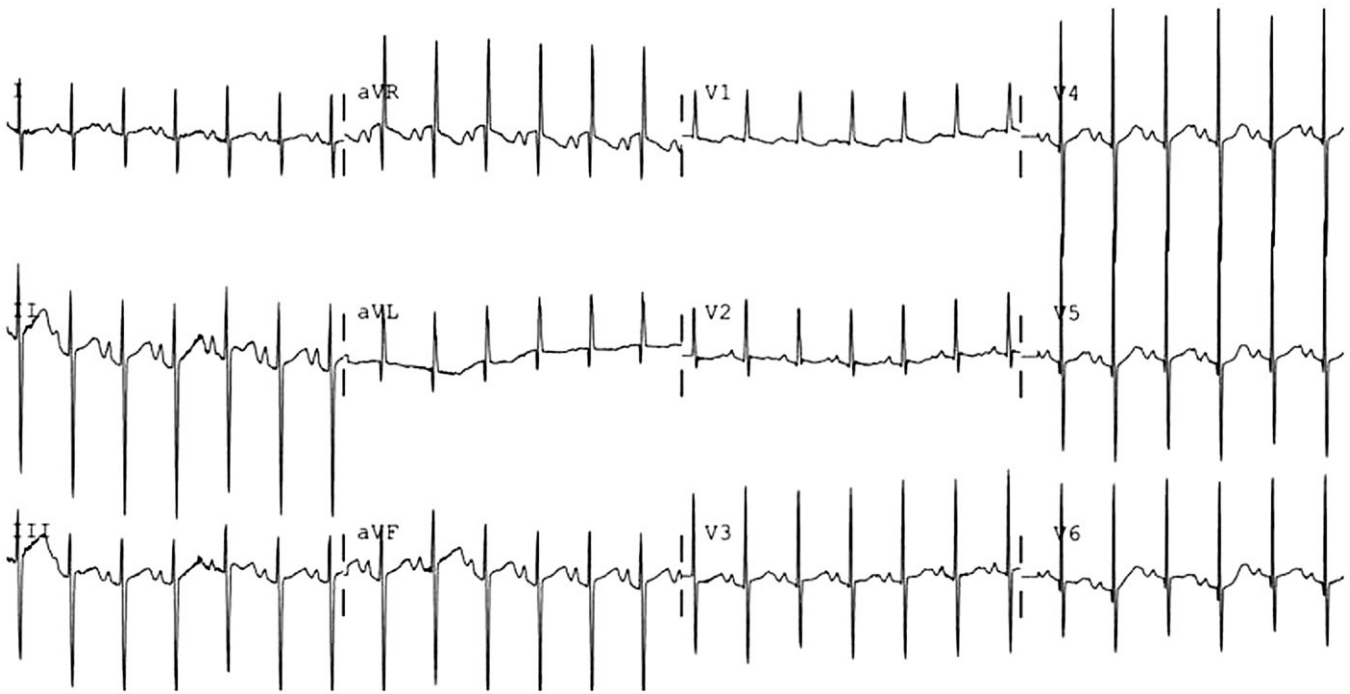
canal defects (Fig. 5) and certainly warrants cardiology consultation. Because the murmur of atrioventricular canal defects can be subtle in the neonatal period, initiation of cardiology evaluation for all infants with Down syndrome is indicated before discharge from the newborn nursery.

Primary rhythm disturbances are uncommon presenting manifestations of CHD. However, certain lesions are associated with arrhythmias easily detected on ECG. Ebstein anomaly of the tricuspid valve (large, dysplastic tricuspid valve leaflets that project inferiorly into the right ventricle, causing an “atrialized” right ventricle) is complicated by preexcitation and other tachyarrhythmias [3]. Varying degrees of AV block extending to complete heart block may be seen in L-transposition of the great arteries, often known as “congenitally corrected” transposition of the great arteries on account of a lack of cyanosis in the newborn period [4]. Heterotaxy syndromes, in which there are abnormalities of cardiac and visceral situs, frequently are accompanied by rhythm disturbances [5].

Myocardial ischemia, as manifested by ST-segment anomalies or by deep Q waves, is fortunately an uncommon finding in pediatric patients. However, several entities should



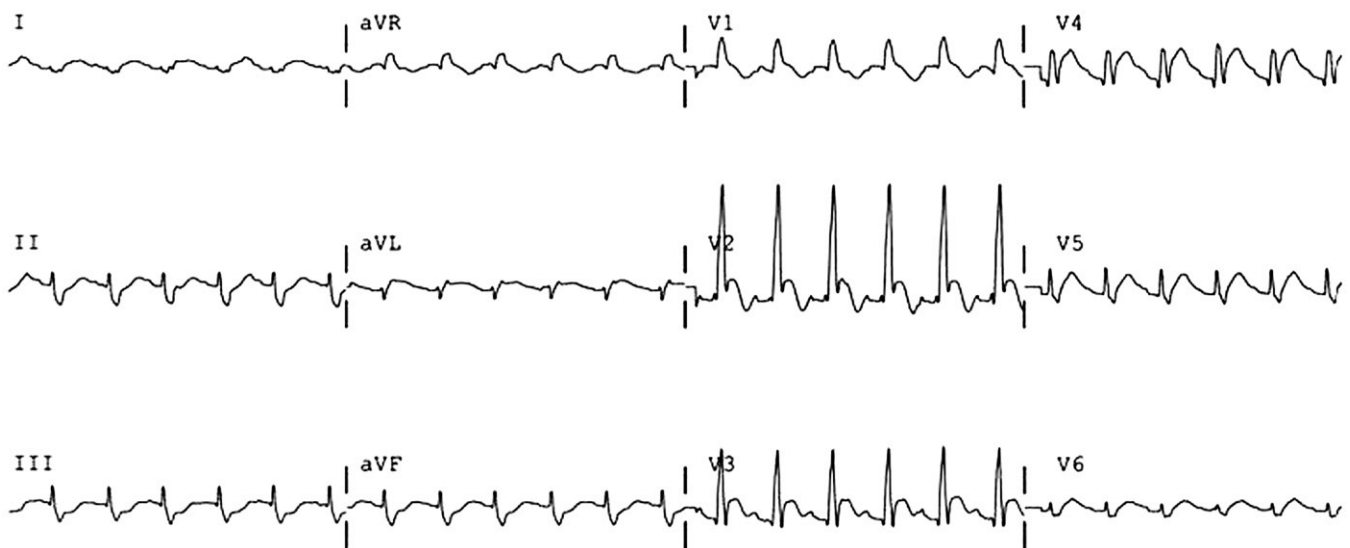
**Fig. 4** Right atrial enlargement in a 5-year-old boy with cardiomyopathy. Note the peaked, large amplitude p waves in lead II.



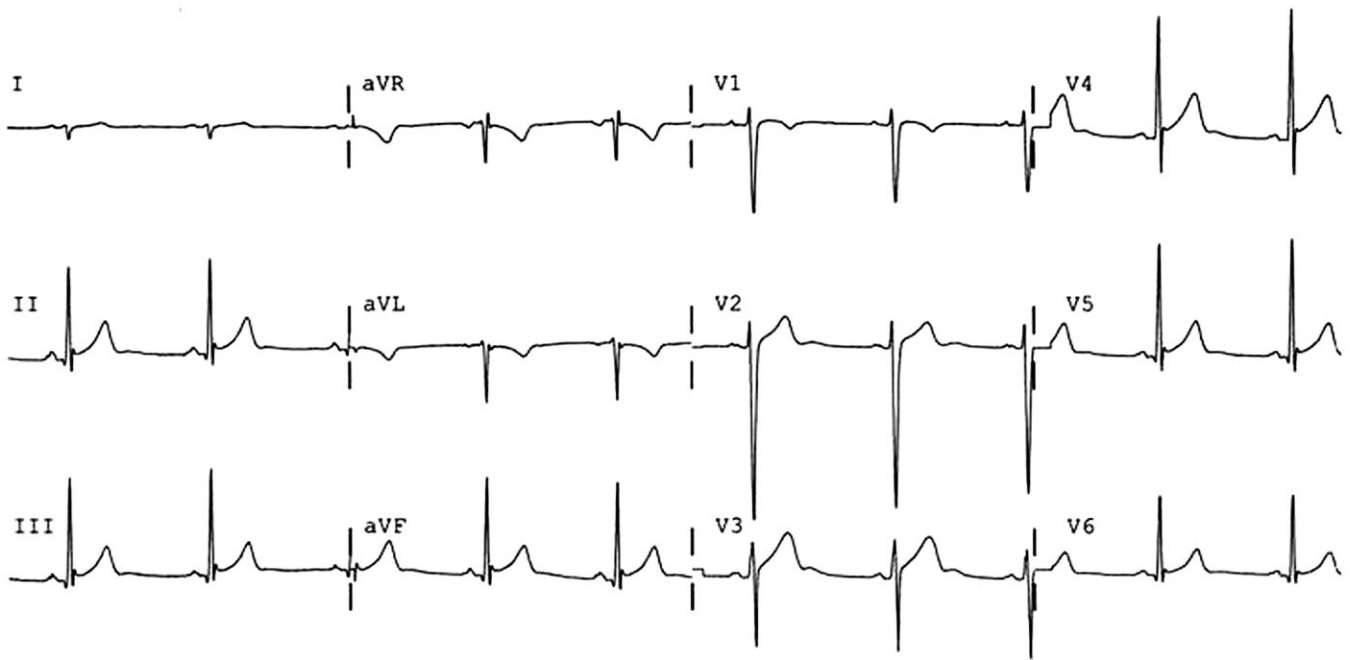
**Fig. 5** ECG in neonate with Down syndrome and atrioventricular canal defect before surgical repair. Mean QRS vector is  $-87^\circ$ , which closely corresponds to “superior axis deviation.”

be considered when such abnormalities are seen on the pediatric ECG. In neonates, diffuse ST-segment elevation may be seen in anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) or, rarely, in hypoplastic left heart syndrome. In ALCAPA, the left coronary artery arises anomalously from the pulmonary artery; as pulmonary arterial pressure falls during the first 6 months of infancy, prograde flow through the left coronary artery ceases and may even reverse [6]. Patients with this condition may present in florid heart failure and be

mistakenly diagnosed as having myocarditis or dilated cardiomyopathy. In hypoplastic left heart syndrome, the coronary arteries are perfused from a hypoplastic, narrow aorta that is susceptible to flow disruption. Finally, children who have undergone orthotopic heart transplantation are at risk of developing transplant allograft vasculopathy [7]. Therefore, an ECG should be performed on all children with heart transplants presenting with acute illness to rule out ischemia. ST-segment elevation is seen in these patients with acute myocardial infarction (Fig. 6).



**Fig. 6** ST elevation (leads V2, V3, and V4) in a 10-year-old boy with cardiac transplant and known coronary allograft vasculopathy. Note the low voltages across all leads. The patient died of overwhelming myocardial infarction.

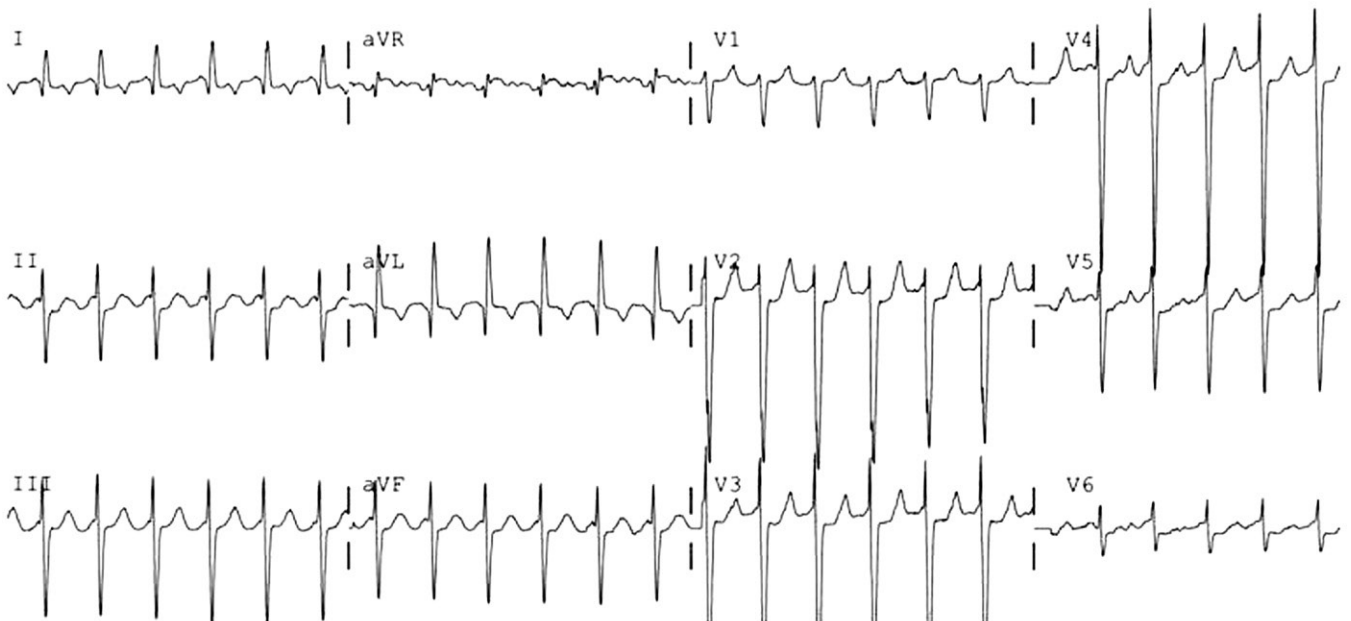


**Fig. 7** BER in a healthy 14-year-old male adolescent. Note the ST-segment elevation, particularly in the precordial leads. The ST elevation is concave, and there are associated tall R waves in the left precordial leads.

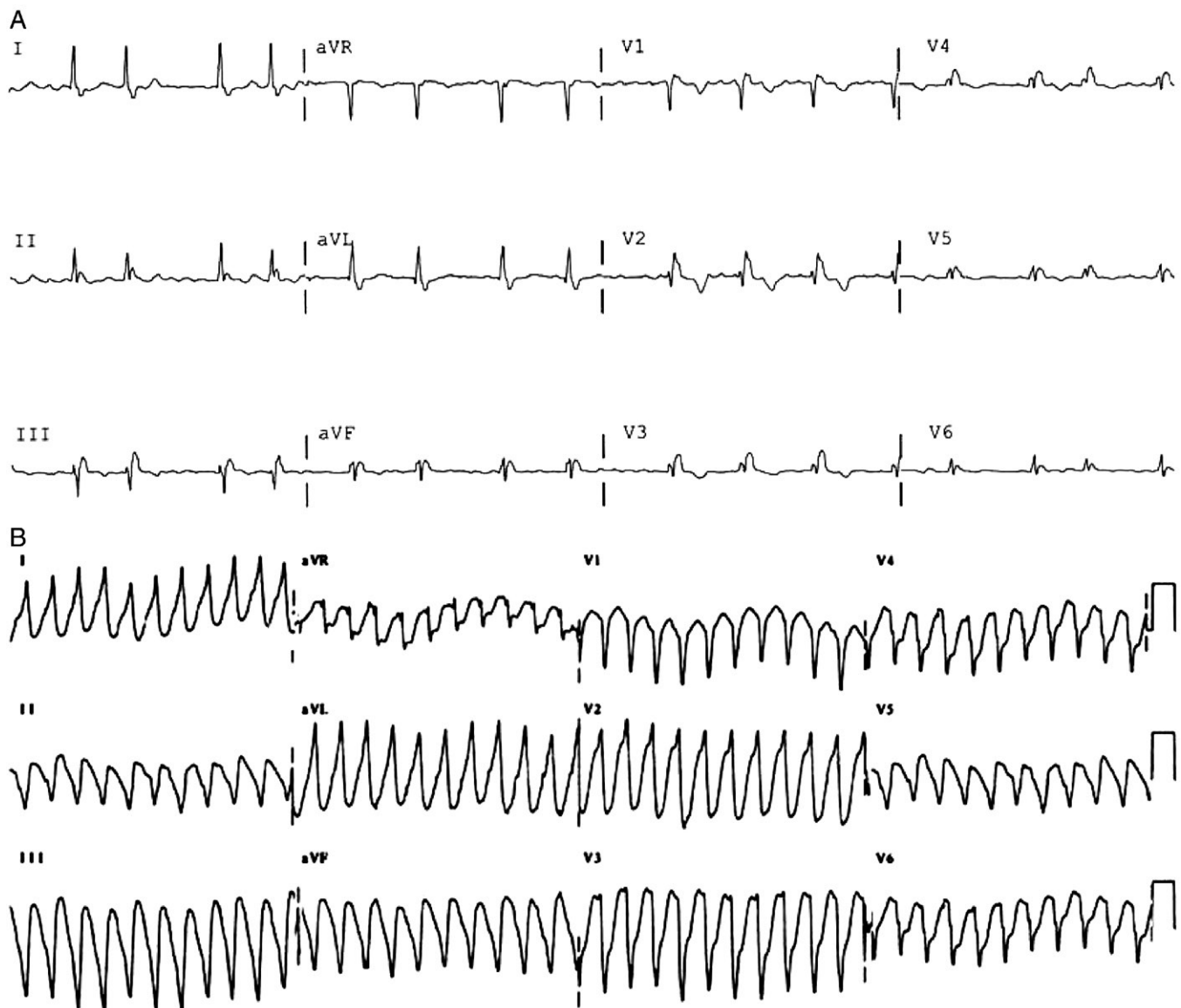
Kawasaki disease, a systemic inflammatory syndrome seen not infrequently in children, may lead to the development of coronary artery aneurysms if the diagnosis and treatment are delayed; subsequent myocardial infarction can occur resulting from thrombosis of the coronary artery aneurysms or stenosis of the coronary artery distal to the aneurysm [8]. Rarely, patients may present several years after an uncategorized febrile illness with symptoms consistent with myocardial infarction due to thrombosis of the coronary

artery aneurysms. In adolescents, cocaine ingestion must also be considered and ruled out when myocardial ischemia is encountered.

ST-segment elevation resulting from ST-segment elevation myocardial infarction and other potentially serious etiologies should be distinguished from benign early repolarization (BER; Fig. 7), an electrocardiographic feature common in younger, healthy patients, particularly male athletes participating in endurance sports. In addition to



**Fig. 8** IART in a 16-year-old male adolescent status post-Fontan procedure. Note its resemblance to atrial flutter.



**Fig. 9** (A) ECG in a 20-year-old woman with repaired tetralogy of Fallot. Note the presence of right bundle branch block and atrial fibrillation. (B) Wide complex tachycardia in the patient from Panel A s/p tetralogy of Fallot repair. Note the AV dissociation, best seen in lead I, strongly suggestive of ventricular tachycardia.

BER, ECG abnormalities, present in up to 40% of highly trained athletes, include increased QRS voltages, diffuse T-wave inversion, and deep Q waves [9].

Patients who have undergone repair for CHD are at increased lifetime risk of arrhythmias, symptomatic and asymptomatic. Several excellent review articles investigate the subject of arrhythmias postsurgery for CHD in detail [10,11]. Many arrhythmias present in the immediate post-operative period, and because these patients are typically monitored in intensive care units, they will not be discussed further. Of more relevance to the acute care provider are disturbances of rhythm that present months to years after surgery. Such disturbances are less common in younger children mainly because newer surgical techniques are

designed to avoid disruption of cardiac conduction systems but are common in adolescents and younger adults.

Atrial flutter is an extremely uncommon rhythm in pediatric patients without a history of CHD [12]. However, surgical repairs involving the atria, particularly those involving the right atrium, have been associated with the development of atrial flutter. Flutterlike atrial tachycardias in the setting of CHD are frequently labeled intra-atrial reentrant tachycardia (IART) due to the slightly slower atrial rate seen in IART and frequent 1:1 AV conduction (Fig. 8). Incisions and suture lines within the right atrium in addition to abnormal hemodynamics are the proposed causative factors. Intra-atrial reentrant tachycardia is classically seen in patients with tricuspid atresia who have undergone a Fontan

procedure as part of staged palliation, particularly those having surgery several decades ago when direct atrio-pulmonary connection was the norm. However, any of the “single ventricle” lesions in which a biventricular circulation cannot be accomplished will require some form of the Fontan procedure and are thus at similar risk of IART. Control of IART is complex and is reviewed in further detail elsewhere [13,14]; patients typically require multiple classes of medications, multiple electrophysiologic procedures, and even perhaps reoperation.

Sinus node dysfunction is a common arrhythmia in the immediate postoperative period for patients with numerous congenital heart defects. In general, the norm is for dysrhythmias of this variety to resolve as the patients exit the acute phase of their hospitalizations [15]. However, permanent AV block is occasionally encountered in patients with surgeries that require manipulations near the AV node and bundle of His; common surgeries include atrioventricular canal repair and older atrial level repairs for transposition of the great arteries (Senning and Mustard procedures).

Q waves representing prior ischemic injury and infarction may be encountered many years after surgery in selected patients. Such infarcts may have occurred as a consequence of the lesion itself or because of the surgical repair; examples in the former group include ALCAPA (described above), whereas a characteristic example of the latter group includes the arterial switch operation for transposition of the great arteries. During this procedure, the coronary arteries are reimplanted onto the aortic root; should there be “kinking” during coronary artery transfer, myocardial ischemia may result.

Ventricular arrhythmias are commonly seen after operations involving incisions made within the ventricular free wall. Repairs of lesions involving RV outflow tract obstruction, classically tetralogy of Fallot, may require ventriculotomies, which may disturb the bundle branches. Patients who have undergone ventriculotomy commonly have varying forms of ventricular conduction disturbance and are known to be prone to ventricular tachycardia and sudden death [16-18]. Because ventricular arrhythmias have been shown to lead to long-term morbidity and mortality, surgical approaches that limit ventriculotomy are being used. In current practice, it remains to be seen whether patients with hypoplastic heart syndrome, many of whom have a RV-pulmonary artery conduit (which requires ventriculotomy) placed in the newborn period, will be at increased risk of ventricular arrhythmias later in childhood and adolescence (Fig. 9A and B).

## 2. Summary

The pediatric ECG is frequently interpreted with apprehension by providers, often for different reasons: the pediatrician examines ECGs with relative rarity, and physicians caring

mainly for adults (family and emergency department physicians) are often unfamiliar with the dynamic nature of the pediatric ECG through childhood and adolescence. A thorough knowledge of the basic age-related changes and norms seen within the pediatric ECG will assist providers in making accurate diagnoses. In addition, familiarity with the pediatric ECG as it applies to CHD will prove valuable for providers who will very likely encounter the ever-growing group of patients who have undergone surgical intervention for CHD.

## References

- [1] Warnes CA, Liberthson R, Danielson Jr GK, et al. Task force 1: the changing profile of congenital heart disease in adult life. *J Am Coll Cardiol* 2001;37:1170-5.
- [2] Friday RY, Ross PA, McDaniel NA. In: Chan TC, Brady WJ, Harrigan RA, et al, editors. *Congenital heart disease*. Philadelphia: Elsevier/Mosby; 2005. p. 238-52.
- [3] Watson H. Natural history of Ebstein's anomaly of tricuspid valve in childhood and adolescence: an international co-operative study of 505 cases. *Br Heart J* 1974;36:417-27.
- [4] Huhta J, Maloney J, Ritter D, et al. Complete atrioventricular block in patients with atrioventricular discordance. *Circulation* 1983;67:1374-7.
- [5] Lin JH, Chang CI, Wang JK, et al. Intrauterine diagnosis of heterotaxy syndrome. *Am Heart J* 2002;143:1002-8.
- [6] Edwards JE. The direction of blood flow in coronary arteries arising from the pulmonary trunk. *Circulation* 1964;29:163-6.
- [7] Pahl E, Naftel DC, Kuhn MA, et al. The impact and outcome of transplant coronary artery disease in a pediatric population: a 9-year multi-institutional study. *J Heart Lung Transplant* 2005;24:645-51.
- [8] Newburger JW, Takahashi M, Gerber MA, et al. Diagnosis, treatment, and long-term management of Kawasaki disease: a statement for health professionals from the Committee on Rheumatic Fever, Endocarditis and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. *Circulation* 2004;110:2747-71.
- [9] Pelliccia A, Maron BJ, Culasso F, et al. Clinical significance of abnormal electrocardiographic patterns in trained athletes. *Circulation* 2000;102:278-84.
- [10] Walsh EP, Cecchin F. Arrhythmias in adult patients with congenital heart disease. *Circulation* 2007;115:534-45.
- [11] Triedman JK. Arrhythmias in adults with congenital heart disease. *Heart* 2002;87:383-9.
- [12] Garson Jr A, Bink-Boelkens M, Hesslein P, et al. Atrial flutter in the young: a collaborative study of 380 cases. *J Am Coll Cardiol* 1985;6:871-8.
- [13] Hebe J. Role of catheter and surgical ablation in congenital heart disease. *Cardiol Clin* 2002;20:469-86.
- [14] Balaji S, Harris L. Atrial arrhythmias in congenital heart disease. *Cardiol Clin* 2002;20:459-68.
- [15] Weindling SN, Saul JP, Gamble WJ, et al. Duration of complete atrioventricular block after congenital heart disease surgery. *Am J Cardiol* 1998;82:525-7.
- [16] Wall K, Oddsson H, Ternstedt BM, et al. Thirty-year electrocardiographic follow-up after repair of tetralogy of Fallot or atrial septal defect. *J Electrocardiol* 2007;40:214-7.
- [17] Gatzoulis MA, Balaji S, Webber SA, et al. Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study. *Lancet* 2000;356:975-81.
- [18] Chandar JS, Wolff GS, Garson Jr A, et al. Ventricular arrhythmias in postoperative tetralogy of Fallot. *Am J Cardiol* 1990;65:655-61.