

Understanding Electrocardiography in Adult Patients With Congenital Heart Disease

A Review

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IMPORTANCE Congenital heart disease in adults is still a relatively new concept for many cardiologists, and the complexity as well as diversity of cardiac phenotypes encountered necessitate that systematic, practical information be available for the nonspecialist. The analysis of the 12-lead electrocardiogram is an invaluable cornerstone in the clinical appraisal of these patients.

OBSERVATIONS Consideration of the main anatomic and pathophysiological aspects of the various congenital heart conditions can shed light on their distinctive electrocardiogram patterns, which are an electrical reflection of intrinsic cardiac anatomy abnormalities, surgical scarring, and progressive cardiac remodeling attributable to hemodynamic perturbations. While congenital heart disease may be diagnosed or suspected on electrocardiogram observation in adults who are previously undiagnosed, specific markers have also been identified to optimize risk stratification in certain defects.

CONCLUSIONS AND RELEVANCE This review outlines that main electrocardiogram patterns in adult patients with congenital heart disease can be appreciated by the understanding of the underlying pathophysiology. Periodic surveillance is of particular importance in this population to unmask early electric signs of disease evolution.

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Recent advances in pediatric cardiology, especially surgical techniques, have resulted in an increasing number of patients with congenital heart disease reaching adulthood, creating a new and steadily growing patient population of adults with congenital heart disease (ACHD).^{1,2} The care for this expanding population, representing more than 1.5 million patients in the US alone, has emerged as a new subspecialty of cardiology.³ However, congenital heart disease in adults is still a relatively new concept for many cardiologists, and the complexity as well as diversity of cardiac phenotypes encountered necessitate that systematic, practical information be available for the nonspecialist. In particular, analysis of the 12-lead electrocardiogram (ECG) remains an invaluable cornerstone in the clinical appraisal of patients with ACHD and often reveals important diagnostic and/or prognostic information.⁴ The ECG patterns in patients with ACHD are an electrical reflection of intrinsic cardiac anatomy abnormalities, surgical scarring, and progressive cardiac remodeling attributable to hemodynamic perturbations. This review outlines the key ECG findings of the most common or distinctive subtypes of ACHD, with brief pathophysiological backgrounds and illustrative examples.

Tetralogy of Fallot

Tetralogy of Fallot (TOF), representing 7% to 10% of all congenital heart diseases, is the most common cyanotic heart defect (found in 1 in 3500 to 4300 adults). First described in 1671 by Niels Stensen,⁵ this condi-

tion became known as *the tetralogy of Fallot* after further description by Etienne-Louis Fallot, a French anatomic pathologist, in 1888.⁶ The 4 characteristic features of TOF are right ventricular (RV) outflow tract obstruction, ventricular septal defect, aortic override, and RV hypertrophy (Figure 1A). In the usual form, without other associated anomalies, modern surgical repair can be performed during the first year of life, sometimes after an initial palliative systemic-pulmonary shunt (Blalock-Taussig anastomosis, usually between the subclavian and the pulmonary artery). Initial corrective surgical approaches (starting in 1955)⁷ involved a large ventriculotomy, wide resection of the pulmonary infundibulum and/or transannular patch, and ventricular septal defect closure. Important pulmonary regurgitation was almost invariably created. Further developments included transatrial and transpulmonary artery approaches, allowing for better preservation of the pulmonary valve function and less scarring on the RV outflow tract musculature, compared with previous access via a ventriculotomy (Figure 1B). Nevertheless, these surgical incisions and patches predispose patients to late development of arrhythmias and sudden cardiac death. The ECG appearances in TOF after surgical repair reflect immediate changes following surgery and subsequent changes associated with progressive RV dilatation and dysfunction.

Rhythm

A baseline sinus rhythm is the rule. While the prevalence of atrial tachyarrhythmias is time dependent, recent data suggest a prevalence of up to 40% by age 45 years. Intra-atrial reentrant tachycar-

dia, mainly right sided, is the most common arrhythmia, although atrial fibrillation is increasing with an aging population.⁸

The P Wave and PR Interval

The P wave is usually normal or peaked, except in cases of severe functional tricuspid regurgitation because of RV dilatation, in which right atrial hypertrophy can be observed. The PR interval is usually normal or mildly increased, but a complete atrioventricular block associated with ventricular septal defect surgical closure may occur.

The QRS and Repolarization

The QRS axis is either normal or has a right deviation attributable to RV hypertrophy and dilatation. An abrupt change from a tall R in V1 to an rS pattern in V2 is often observed, possibly reflecting loss of myocardium over the RV outflow. A right bundle branch block is almost universal, resulting from surgical injury to the right bundle branch and/or RV hypertrophy and dilatation. The right bundle block morphology is often atypical, with variable QRS prolongation in different leads and a maximal duration in right-sided precordial leads, which reflect slow conduction around the ventriculotomy incision or a patch with (most of the time) a focal conduction block rather than right bundle branch block in the trunk. While multiple criteria mostly derived from the amplitude of R and S waves in leads V1 and V6 are proposed for RV hypertrophy diagnosis, to our knowledge, no specific criteria have been proposed for RV hypertrophy in patients with right bundle branch block,⁹ although tall R waves in right precordial leads are suggestive. The accuracy of electrographic criteria for RV hypertrophy remains relatively low in these patients, and a multimodal evaluation including cardiac imaging is helpful.

Repolarization abnormalities with diffuse ST and T wave changes are frequently observed. Increased dispersion of QT or other repolarization intervals have been reported¹⁰ and reflect a potentially arrhythmogenic substrate attributable to myocardial fiber derangement. This likely contributes to the development of ventricular arrhythmias triggered by increased heterogeneity in myocardial repolarization.

Risk Stratification

The main ECG risk factor for ventricular arrhythmias in adults with repaired TOF is QRS prolongation and in particular a QRS duration of 180 ms or more.¹¹ Also, QRS fragmentation, defined as 3 or more R waves or notches in the R/S complex in 2 or more contiguous leads (or 2 or more in the absence of bundle block branch), has been more recently associated with outcomes with TOF, but its value for prognosticating sudden death specifically remains sparsely assessed.¹² The QRS change over time is also of particular interest, because progressive broadening has been associated with an increased risk of sudden cardiac death. However, risk stratification remains complex in TOF, and other markers, such as nonsustained ventricular tachycardia and some non-ECG parameters, should be considered in identifying patients at risk.^{13,14}

Ebstein Anomaly

Ebstein anomaly (EA) accounts for less than 1% of all congenital heart disease, but milder forms may go undiagnosed.¹⁵ First described on autopsy by Wilhelm Ebstein in 1866,¹⁶ EA is characterized by a mal-

formed and apically displaced tricuspid valve. A portion of the RV is atrialized (ie, functionally integrated within the right atrium) (Figure 1D). The consequence of EA is tricuspid regurgitation of varying severity (depending on the degree of displacement and the functional status of the tricuspid valve leaflets), resulting in right atrial and RV enlargement. An interatrial communication (atrial septal defect or patent foramen ovale) is present in more than 80% of patients.¹⁵ Furthermore, there is an adverse ventricular-ventricular interaction, which with time leads to RV dysfunction. When possible, surgical repair vs valve replacement should be considered. Different techniques of tricuspid valvuloplasty involving various modifications of plication or resection of the atrialized ventricle have been proposed (Figure 1E).

Rhythm

While most patients with EA present with a sinus rhythm, atrial arrhythmias are frequent and correspond to right atria dilatation. Supraventricular tachyarrhythmias are the most common mode of presentation in adolescents and adults.¹⁵

The P Wave and PR Interval

Large and tall P waves (called *Himalayan P waves*) are common as a result of right atrial dilatation. First-degree AV block is frequent, reflecting an intra-atrial conduction delay. Short PR interval and delta waves can also be seen, since accessory pathways are reported in 20% of the cases. These accessory pathways are usually right-sided (around the abnormal tricuspid annulus) and often multiple.

The QRS and Repolarization

Incomplete or complete atypical right bundle branch block is usually observed in EA. The terminal parts of the QRS complexes are fragmented and multiphasic, and this results from abnormal activation of the atrialized RV.¹⁷ Low-amplitude R waves are characteristically seen in right precordial leads and reflect the diminutive functional RV.

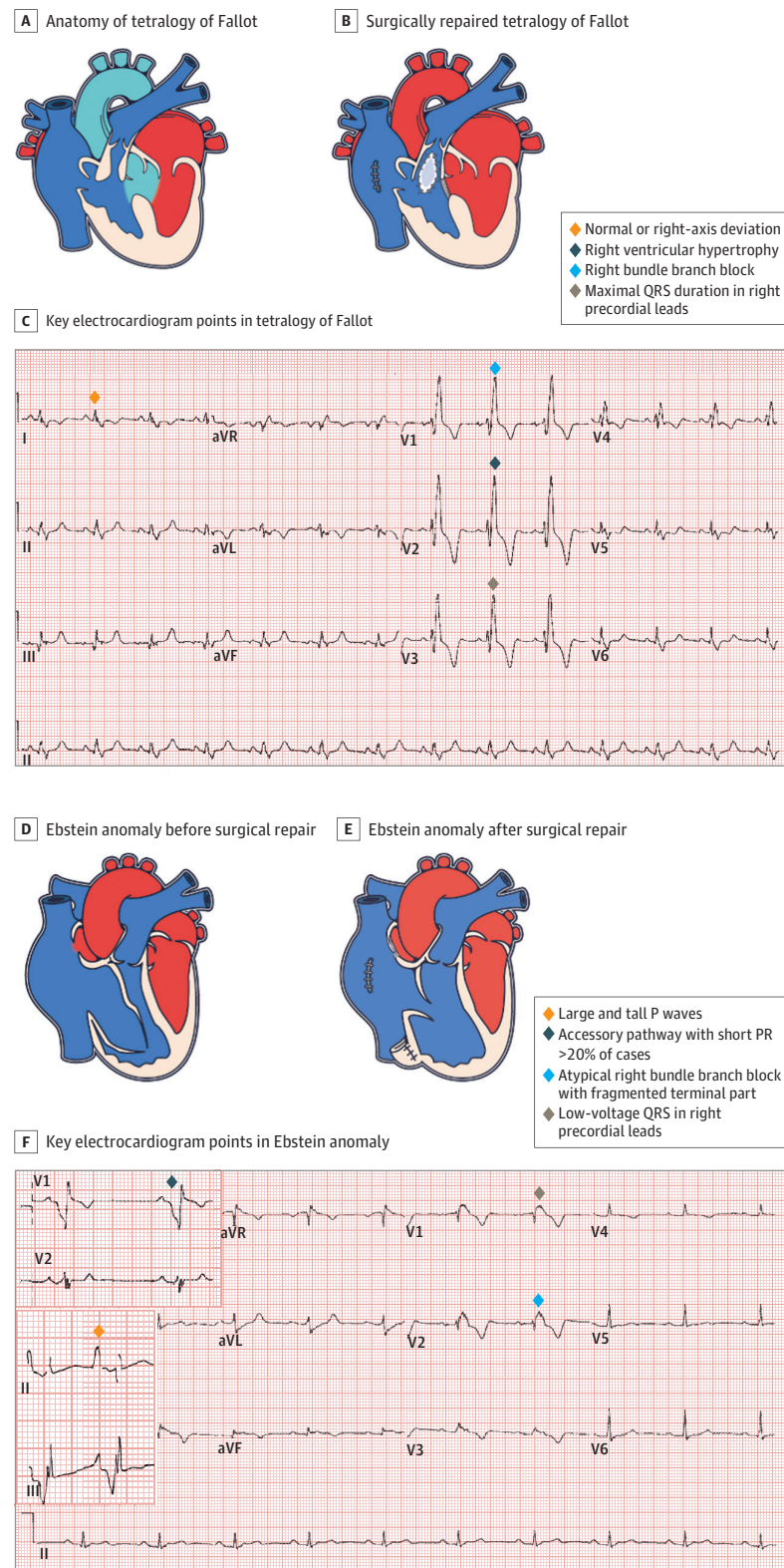
Risk Stratification

Atrial arrhythmias and QRS broadening or fragmentation have been associated with ventricular arrhythmic events in EA.^{18,19} Also, sudden cardiac death can occur because of rapid conduction of atrial fibrillation or flutter to the ventricles via an accessory pathway. Catheter ablation of high-risk or multiple accessory pathways should be performed, and systematic electrophysiological evaluation has been proposed by some teams before surgical repair.²⁰ Ablation may be rendered challenging by the presence of multiple pathways (in up to 50% of cases²¹) and complex insertion patterns along the abnormal tricuspid annulus, with difficulty identifying the true atrioventricular (AV) groove.

Transposition of the Great Arteries With Intra-atrial Baffle Repair (Mustard and Senning Procedures)

D-Transposition of the great arteries (D-TGA; also known as *complete transposition*) accounts for 5% of all congenital cardiac malformations²² and is characterized by the aorta originating from the RV and the pulmonary trunk originating from the left ventricle (Figure 2A). This anomaly is incompatible with survival in the

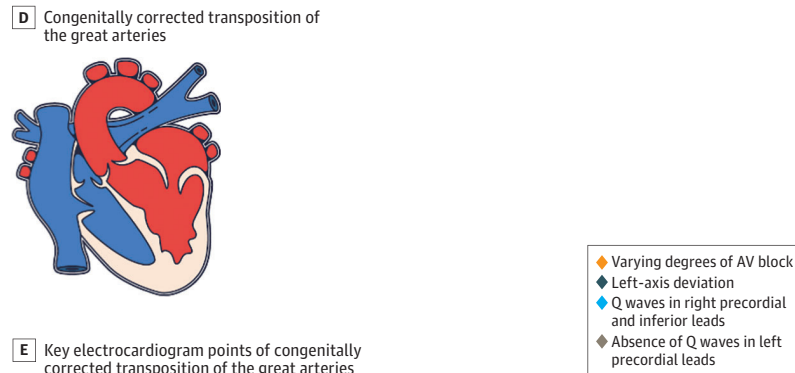
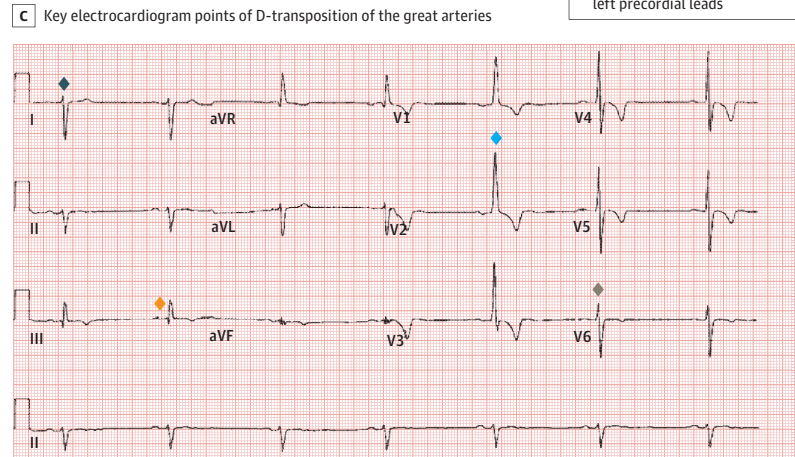
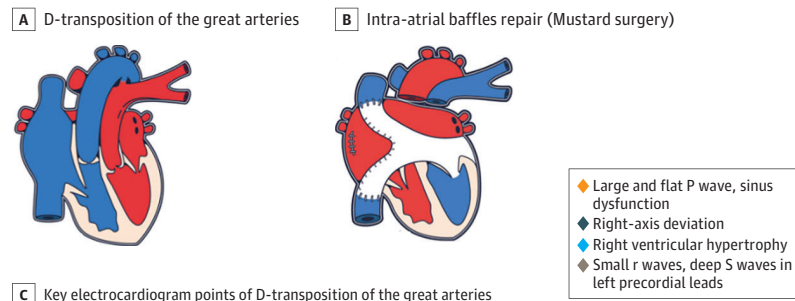
Figure 1. Tetralogy of Fallot and Ebstein Anomaly



absence of some form of communication between the systemic and pulmonary circulations. Immediate management of TGA in

infancy involves preservation of the open ductus arteriosus or the creation of an atrial septal defect by balloon atrial septostomy

Figure 2. D-Transposition of the Great Arteries and Congenitally Corrected Transposition of the Great Arteries

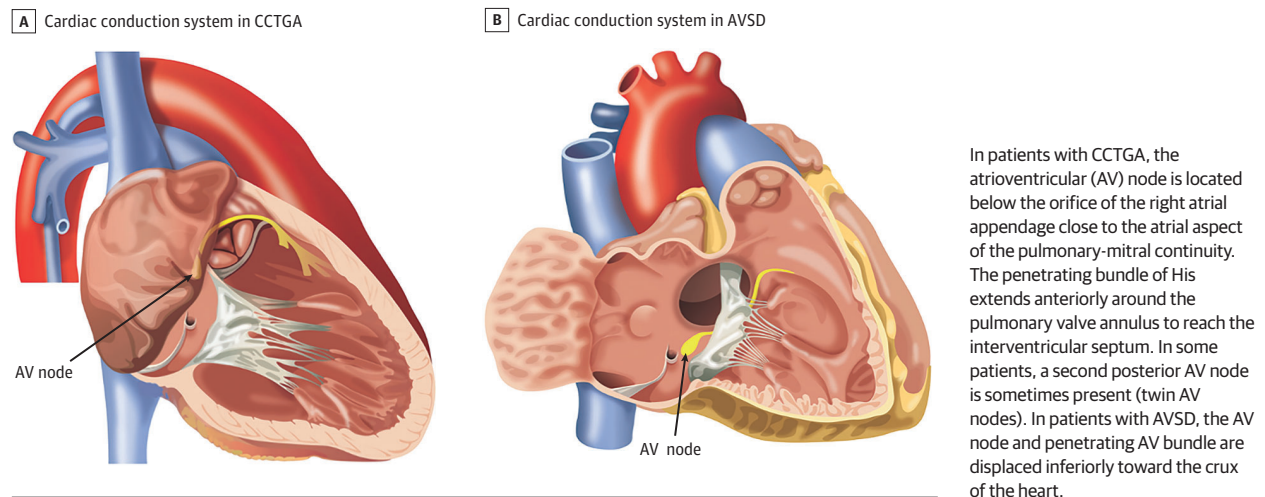


AV indicates atrioventricular.

(Rashkind maneuver) to maintain or create a communication between the 2 separate circulations. Currently, an arterial switch

operation is performed during the first days of life (connecting the aorta to the left ventricle and the pulmonary trunk to the RV, with

Figure 3. Cardiac Conduction System in Congenitally Corrected Transposition of the Great Arteries (CCTGA) and Atrioventricular Septal Defects (AVSD)



corresponding coronary reimplantation) with excellent outcomes. However, before the mid-1980s,²³ this anatomic repair was not an option; instead, a physiologic repair was used, which involved the construction of a baffle within the atria to direct systemic venous blood across the mitral valve into the left ventricle and the pulmonary venous blood across the tricuspid valve into the RV (atrial switch). The Senning operation²⁴ was first introduced in 1959 to redirect blood via a relatively complex construction of autologous atrial tissue, whereas in 1963, William Mustard²⁵ proposed an alternative technique that used a pericardial patch (Figure 2B). Thus, a physiologic circulation was restored, although the RV continued to support the systemic circulation. Although arterial switch surgery is currently the procedure of choice, most adults with D-TGA at present have had intra-atrial baffle repairs and present with distinctive ECG features.

Rhythm

Large and complex atrial surgical scars cause frequent sinus node dysfunction and intra-atrial conduction disturbance. Ectopic atrial and junctional rhythms are commonly observed, and up to one-quarter of these patients will require pacemaker implantation by age 25 years.^{26,27} A high prevalence of intra-atrial reentrant tachycardia (atrial flutter) is also reported, with multiple reentrant circuits.^{28,29}

The P Wave and PR Interval

The P wave is usually large and flat, with low voltage because of extensive atrial scar tissue. While AV node function is preserved most of the time in patients without coexisting congenital heart defects, intra-atrial conduction delay may result in a prolonged PR interval.³⁰

The QRS and Repolarization

Since the RV remains the systemic ventricle after atrial switch, RV hypertrophy, right-axis deviation, and a right bundle block pattern are the norm. The RV overload is characterized by a high voltage in right precordial leads and a late transition of the QRS complex after V3. The left ventricle in the subpulmonary position is not well developed, which is reflected by small r waves and deep S waves over the left precordial leads.⁴

Risk Stratification

Atrial arrhythmias are common and associated with sudden cardiac death after atrial switch procedures.³¹ Atrial flutter with 1:1 conduction is frequent, because patients with Senning or Mustard repairs have initially normal atrioventricular nodal conduction and a slower flutter cycle length attributable to surgical atrial scars. Rapid ventricular response to supraventricular arrhythmias can cause clinically significant deterioration in systemic RV function and compromise hemodynamics and/or trigger ventricular arrhythmias.³² Other ECG criteria have not been found to reproducibly prognosticate sudden cardiac death. Primary ventricular arrhythmias appear to be uncommon, unless there is coexisting severe RV dysfunction.²⁹

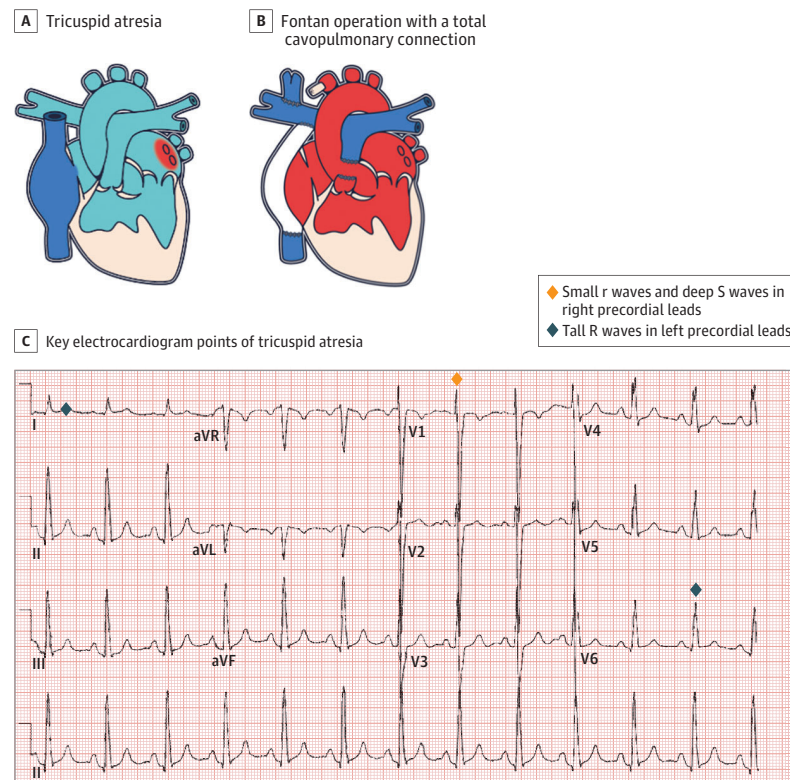
Congenitally Corrected Transposition of the Great Arteries

Congenitally corrected transposition of the great arteries (CCTGA; also named *L-transposition of the great arteries*) is a rare defect (<1% of congenital heart disease), characterized by atrioventricular and ventriculoarterial discordance. Ventricles and their attached valves are reversed, with a subaortic RV and a subpulmonary left ventricle (Figure 2D). Patients with CCTGA may remain asymptomatic and be diagnosed later in life, although associated anomalies (eg, severe tricuspid regurgitation, ventricular septal defect, and/or subpulmonary outflow obstruction) can be responsible for a much earlier presentation. The systemic atrioventricular valve is sometimes of Ebstein type, with insertion abnormalities that account for a variable degree of intrinsic regurgitation. In the absence of significant associated anomalies, surgery or other intervention is rarely required, and the course of this condition mainly depends on the function of the morphological RV that supports the systemic circulation.

Rhythm

The AV node and His bundle in CCTGA have an unusual position, which is anterior and slightly more lateral than typical (Figure 3A).

Figure 4. Tricuspid Atresia and Fontan Operation With a Total Cavopulmonary Connection



The AV conduction system is particularly fragile because of the abnormal course, with a high rate of complete AV block, estimated to occur in 2% to 3% of patients annually.³³ Significant systemic tricuspid regurgitation may promote atrial enlargement and atrial arrhythmias.

The P Wave and PR Interval

The P wave morphology and axis are usually normal. Large or tall P waves may be observed in cases of severe systemic atrioventricular valve regurgitation. The PR interval is frequently prolonged, with varying degrees of AV block. Ventricular pre-excitation with a short PR interval may also be present, since atrioventricular accessory pathways are relatively frequent around the Ebstein-type valve.

The QRS and Repolarization

Ventricles and associated bundle branches are reversed, causing septal activation to occur from right to left. This contributes to the pathognomonic CCTGA ECG pattern: Q waves in right precordial leads and absent Q waves in left precordial leads. Left-axis deviation is the rule, and QRS duration is usually normal in the absence of advanced systemic RV dilation and dysfunction.³⁴

Risk Stratification

Although reports on long-term outcomes of adults with CCTGA have emphasized sudden cardiac death as an important cause of mortality, the small number of patients in most reported series has precluded identification of reliable risk factors. Attention has primarily focused on hemodynamic parameters (eg, progressive systemic right

ventricular dysfunction and/or atrioventricular valve regurgitation), as opposed to ECG parameters.³⁵

Univentricular Hearts and Fontan Surgery: Example of Tricuspid Atresia

The univentricular heart represents a wide range of heterogeneous congenital heart defects characterized by a single functional ventricle and a second rudimentary or hypoplastic ventricular chamber. Although the most common form of univentricular heart is hypoplastic left-heart syndrome, the most prevalent defect in adults is currently tricuspid atresia (absence of a tricuspid orifice, a hypoplastic RV, and interventricular communication) (Figure 4A). Early management includes atrial septostomy (Rashkind maneuver) and/or a systemic-pulmonary shunt to maintain pulmonary blood flow. An atriopulmonary anastomosis (first described by Francis Fontan in 1971³⁶) or most recently total cavopulmonary anastomosis is then performed between the ages of 1 and 4 years, resulting in separated pulmonary and systemic circulations. The classic Fontan procedure involved an anastomosis between the right atrial appendage and the main pulmonary artery, but with time leads to major right atrial dilatation, with consequent arrhythmias, thrombosis, and right heart failure.³⁶ The modified Fontan approach now involves an anastomosis between the superior vena cava and the right pulmonary artery (Glenn anastomosis) and either an intra-atrial lateral tunnel or, more recently, an extra cardiac conduit connecting the inferior vena cava to the pulmonary artery (Figure 4B).³⁷

Rhythm

Atrial arrhythmias, which are mostly attributable to macroreentrant circuits, depend on the type of surgical route but have been reported in more than 50% of patients during long-term follow-up.³⁸ These arrhythmias often respond poorly to antiarrhythmic pharmacological therapy and may result in rapid hemodynamic deterioration and heart failure. Sinus node dysfunction is also frequent in these patients and increases with time.³⁹

The P Wave and PR Interval

The P waves are generally tall and broad in a classic Fontan configuration but depends on underlying phenotype and type of surgical repair, with P-wave duration shorter after extra cardiac conduit compared with an intra-atrial lateral tunnel or atriopulmonary connection.⁴⁰ The PR interval is usually normal in tricuspid atresia, whereas PR prolongation or a higher degree of AV block may be observed in other subtypes of single-ventricle physiology, particularly in hearts with AV discordance or atrioventricular septal defects (AVSD).

The QRS and Repolarization

The RV is diminutive in tricuspid atresia. Therefore, left ventricular preponderance is reflected in frequent left-axis deviation, small r waves and deep S waves in right precordial leads, and tall R waves in left precordial leads.⁴¹

Risk Stratification

Atrial arrhythmias in so-called univentricular hearts may be associated with substantial morbidity and increased risk of sudden cardiac death because rapid hemodynamic deterioration can ensue.⁴² Catheter ablation is often required, but management remains challenging because multiple new circuits can develop over time.

Atrioventricular Septal Defect

Atrioventricular septal defect, also referred to as an *atrioventricular canal* or *endocardial cushion defect*, covers a spectrum of congenital heart malformations characterized by a common AV junction coexisting with deficient AV septation. In partial AVSD (ostium primum atrial septal defect [ASD] and so-called cleft left AV valve), there are separate atrioventricular valve orifices despite a common junction, while in complete AVSD, the valve itself is also shared (up to 5 leaflets) with a variable deficiency of the inlet ventricular septum (Figure 5A).⁴³ Atrioventricular septal defect is strongly associated with Down syndrome. Partial defects or surgically repaired complete AVSD are the most common presentations in adulthood. Complete AVSD results in significant interatrial and interventricular communication, inducing both right atrial and ventricular overload and congestive heart failure. Surgical repair, which consists of closing intracardiac communications with patches and construction of 2 separate and competent AV valves, is usually performed before the sixth month of life to prevent the development of pulmonary vascular disease (Figure 5B).

Rhythm

A sinus rhythm is the rule. Atrial arrhythmias are relatively rare but may occur, mainly in enlarged atria, because of late repair or around surgical scars.

The P Wave and PR Interval

The P wave is usually normal, except in cases of right atrial dilatation, because of a persistent or late closing of the defect. The PR interval is commonly prolonged, and first-degree AV block is observed in more than half of patients with AVSD, mainly caused by intra-atrial conduction delay.⁴⁴ Complete AV block can occur at the time of surgery.

The QRS and Repolarization

The posteroinferior displacement of the AV node and the relative hypoplasia of the left anterior fascicle result in an early impulse propagation to the posterior aspect of the ventricular septum and left ventricle, giving rise to the distinctive AVSD ECG pattern: superior QRS axis (leftward or extremely rightward), predominant S waves in inferior leads, and R waves in aVL and aVR (Figure 3B).⁴⁵ Incomplete right bundle branch block may also be observed and could result from delayed Purkinje conduction, as well as from conduction over the longer-than-normal right bundle branch that arises from the displaced His bundle.⁴⁶

Risk Stratification

Low incidence of ventricular arrhythmias and sudden cardiac death has been reported in long-term follow-up of patients with AVSD. No ECG marker has been specifically identified.⁴⁷

Atrial Septal Defect

Atrial septal defect accounts for 30% of congenital heart disease⁴⁸ detected in adulthood. Three major types of interatrial communications are described: ostium secundum, ostium primum, and sinus venosus defects. The ostium secundum is the most prevalent and involves the region of the fossa ovalis. The ostium primum defect is part of the spectrum of AVSD, associated with a trileaflet (cleft) left AV valve, often misnamed as a mitral valve. The sinus venosus defect is usually located at the junction of the right atrium and superior vena cava and almost always associated with partial anomalous pulmonary venous return (Figure 5D). The magnitude of the left-to-right shunt depends on the ASD size and the relative diastolic filling properties of the left and right ventricles. A sizeable defect results in dilation of the right heart chambers and may cause pulmonary hypertension, right heart failure, supraventricular tachycardia, and paradoxical systemic embolism. Patients with symptomatic or hemodynamically significant ASDs should be offered elective closure, irrespective of age. A percutaneous approach is usually preferred, but surgery is required for patients with ostium primum and sinus venosus ASDs or patients with ostium secundum defects when anatomy is unsuitable for device closure.

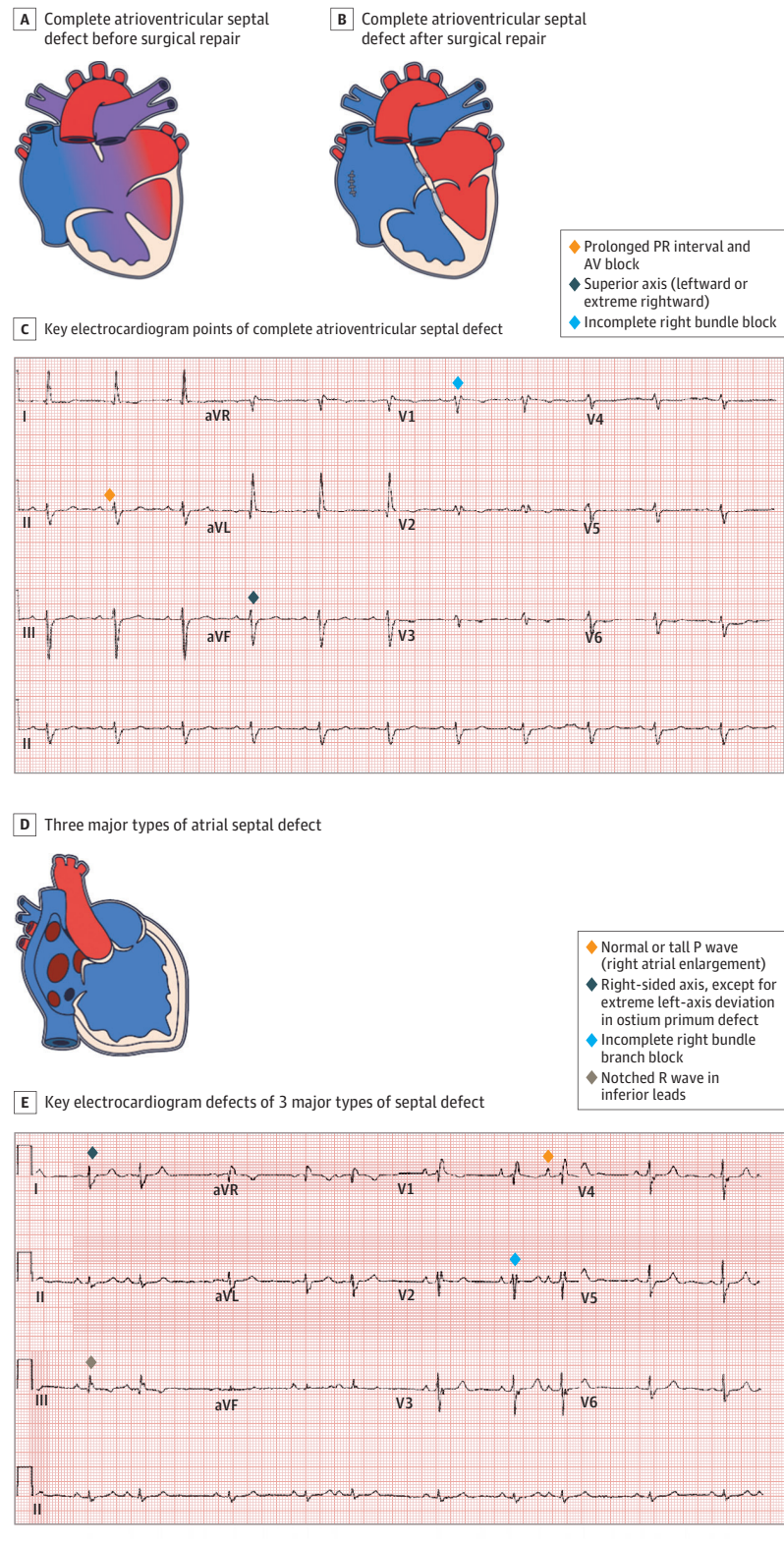
Rhythm

Most patients with ASDs present with a normal sinus rhythm. Late ASD repair is associated with an increased risk of atrial arrhythmia, mainly atrial fibrillation and intra-atrial reentrant tachycardia (ie, atrial flutter or an incisional macroreentrant circuit).⁴⁹

The P Wave and PR Interval

The P wave is usually normal, but right atrial overload is observed in the case of large or late repair of an ASD. A junctional or low atrial rhythm with inverted P waves in the inferior leads suggests an ab-

Figure 5. Complete Atrioventricular Septal Defect and Atrial Septal Defects



Because a sagittal plane is used to visualize main types of atrial septal defect in D, only the right ventricle is depicted. AV indicates atrioventricular.

sent or deficient sinus node, as may be seen in sinus venosus defects after surgery.⁵⁰ First-degree AV block suggests an ostium primum defect and may also be seen in older patients with ostium secundum ASDs.

The QRS and Repolarization

The RV overload manifests as an unusual form of incomplete right bundle branch block, with a broader and slurred terminal r' wave. An rSR' or rSr' pattern is a classic observation in right precordial

leads. Complete right bundle branch block can occur in older patients. A notch of the terminal R wave in inferior leads has also been reported in 75% of patients with ASDs, with a high specificity.⁵¹ The electrical axis is typically normal or right sided, but extreme left-axis deviation is noted in ostium primum defects, since conduction bundles are usually shifted downward by the septal defect with a hypoplastic left anterior fascicle.

Risk Stratification

Long-term follow-up studies have reported a very low risk of sudden cardiac death after ASD repair, precluding identification of reliable ECG risk markers.^{52,53} Atrial arrhythmias have been associated with a higher overall mortality, particularly in men.⁵⁴

Conclusions

Consideration of the main anatomic and pathophysiological aspects of the various ACHD conditions can shed light on their distinctive ECG patterns. While congenital heart diseases may be diagnosed or suspected by ECG observation in adults with no previous diagnosis, specific markers have also been identified to optimize risk stratification in certain defects, and their periodic surveillance is of clinical relevance. The understanding of the main ECG features is of particular importance, not only for ACHD specialists, but also for general cardiologists, since the population of patients with ACHD is continuously expanding.

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