Clinical Use of Electrocardiography in Adults With Congenital Heart Disease

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The prevalence of adult congenital heart disease (ACHD) has risen markedly over the past 2 decades, with the number of adults now rivaling the number of children with severe defects. This is, perhaps, not surprising given that current care allows nearly 90% of infants born with heart defects to thrive into their adult years. This remarkable triumph is tempered, however, by the realization that early interventions were reparative and not curative. Numerous complications may surface years after uneventful childhood courses, justifying vigilant clinical follow-up throughout adulthood. The 12-lead ECG remains an invaluable cornerstone in the clinical appraisal of adults with congenital heart disease that, in certain circumstances, provides diagnostic and/or prognostic information. The present review imparts a clinical perspective to ECG interpretation in ACHD, emphasizing practical and pathogenomic findings in the more frequently encountered congenital defects in adults. Anatomic features of the conduction system relevant to ECG findings in ACHD are summarized, including variations in the location of the sinus node, atrioventricular (AV) node, and His-Purkinje system. Thereafter, pertinent ECG features are highlighted for common subtypes of ACHD (Table). Examples are provided throughout for illustration.

Anatomy of the Conduction System in ACHD

Sinus Node

In the morphologically normal heart, a crescent-shaped sinus node is characteristically located epicardially along the lateral aspect of the superior cavoatrial junction. It generates a P-wave axis typically between 15° and 75°. Most patients with ACHD have normally positioned atrial chambers, called atrial situs solitus, with normal sinus node location. The position of the sinus node may, however, vary with the atrial chambers and their appendages.

Juxtaposition of the Atrial Appendages

In juxtaposition of the atrial appendages, both appendages are on the same side of the arterial pedicle rather than each being ipsilateral to its respective atrium. Left juxtaposition, with left-sided atrial appendages, frequently accompanies tricuspid atresia and has been associated with abnormal ventriculoarterial connections (eg, discordant or double-outlet right ventricle). The sinus node is displaced anterior and inferiorly, below the crista terminalis. Right juxtaposition is far less common and is not associated with a malposed sinus node.

Situs Inversus

In atrial situs inversus, the atria are positioned in a mirror-image fashion, with the right atrium and its sinus node on the individual’s left side. Consequently, the P-wave axis is between 105° and 165°, is often negative in lead I, and most positive in lead III.

Heterotaxy Syndromes

Heterotaxy syndromes are disorders of lateralization whereby the arrangement of abdominal and thoracic viscera differs from situs solitus and inversus. Despite much variability, these usually severe defects are characterized as either right (asplenia syndrome) or left (polysplenia syndrome) atrial isomerism. Patients with right atrial isomerism often have 2 distinct sinus nodes at the junction of right- and left-sided superior vena cavae with atrial chambers. The P-wave axis may fluctuate between governing sinus nodes (Figure 1). In contrast, in left atrial isomerism, sinus nodes are either absent or hypoplastic and posteroinferiorly displaced, far from the superior vena cava orifice. As a result, slow atrial rates and junctional escape rhythms are common.

AV Node and His-Purkinje System

In normal hearts, the AV node is located at the apex of Koch’s triangle, delimited by the septal leaflet of the tricuspid valve, tendon of Todaro (which extends from the eustachian valve to the central fibrous body), and coronary sinus ostium. The His bundle extends from the AV node as the only pathway of myocardial continuity between atria and ventricles. Displacement of the AV conduction system generally accompanies disorders with misaligned atrial and ventricular septae, discordant AV arrangements, and single ventricles.

Atrioventricular Canal Defect

In AV canal defects (AVCDs), the compact AV node is inferiorly and posteriorly displaced outside Koch’s triangle adjacent to where posterior rims of atrial and ventricular septae unite. The His bundle extends along the lower rim of the ventricular septum. This inferior course and relative...
hypoplasia of the left anterior hemisecfacle gives rise to the
superior QRS axis typical of AVCD.\(^9\)

**Congenitally Corrected Transposition of the Great Arteries**

In congenitally corrected transposition of the great arteries (TGA), AV and ventriculoarterial discordance coexist with a
normal atrial situs. The right ventricle is subaortic and
leftward. The AV node is displaced outside of Koch’s
triangle, anterior and slightly more laterally.\(^10\) An elongated
His bundle extends toward the site of fibrous continuity
between the right-sided mitral valve and pulmonary artery. It
courses across the anterior rim of the pulmonary valve and
continues along the superior border of a ventricular septal
defect (VSD), if present.\(^10\)

**Tricuspid Atresia**

Patients with tricuspid atresia have an imperforated fibrous
AV connection. Anterior to the coronary sinus ostium, a

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**Table. Typical ECG Features in Common Forms of ACHD**

<table>
<thead>
<tr>
<th>Congenital Diagnosis</th>
<th>Rhythm</th>
<th>PR Interval</th>
<th>QRS Axis</th>
<th>QRS Configuration</th>
<th>Atrial Enlargement</th>
<th>Ventricular Hypertrophy</th>
<th>Particularities</th>
</tr>
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<tbody>
<tr>
<td>Secundum atrial septal defect</td>
<td>NSR; ↑ IART/AF with age</td>
<td>1° AVB 6% to 10%</td>
<td>0° to 180°; RAD; LAD in Holt-Oram or LAHb</td>
<td>rs’r or rs’r with RBBB→rBBBc</td>
<td>RAE 35%</td>
<td>Uncommon</td>
<td>“Crochetage” phenomenon</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>NSR; PVCs</td>
<td>Normal or mild ↑; 1° AVB 10%</td>
<td>RAD with BH; LAD 3% to 15%</td>
<td>Normal or rs’; possible RBBB</td>
<td>Possible LAE</td>
<td>BWV 23% to 61%; RSV with Eisenmenger</td>
<td>Katz-Wachtel phenomenon</td>
</tr>
<tr>
<td>AV canal defect</td>
<td>NSR; PVCs 30%</td>
<td>1° AVB &gt;50%</td>
<td>Moderate to extreme LAD; normal with atypical</td>
<td>rs’r or rs’r</td>
<td>Possible LAE</td>
<td>Uncommon in partial; BHV in complete; RSV with Eisenmenger</td>
<td>Interposterolateral displaced AVN</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>NSR; ↑ IART/AF with age</td>
<td>↑ PR 10% to 20%</td>
<td>Normal</td>
<td>Deep S V₃, tall R V₅ and V₆</td>
<td>LAE with moderate PDA</td>
<td>Uncommon</td>
<td>Often either clinically silent or Eisenmenger</td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
<td>NSR</td>
<td>Normal</td>
<td>Normal if mild; RAD with moderate/severe</td>
<td>Normal; or rs’r; R’ increases with severity</td>
<td>Possible LAE</td>
<td>RSV; severity correlates with R:S in V₃ and V₆</td>
<td>Axis deviation correlates with RVP</td>
</tr>
<tr>
<td>Aortic coarctation</td>
<td>NSR</td>
<td>Normal</td>
<td>Normal or LAD</td>
<td>Normal</td>
<td>Possible LAE</td>
<td>LVH, especially by voltage criteria</td>
<td>Persistent RSV rare beyond infancy</td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
<td>NSR; possible EAR, SVT; AF/IART 40%</td>
<td>1° AVB common; short if WPW</td>
<td>Normal or LAD</td>
<td>Low-amplitude multiphasic atrialypical RBBB</td>
<td>RAH with Himalayan P waves</td>
<td>Possible RV if RVOT obstruction or PHT</td>
<td>QRS duration ≥ 0.08 sec prognostic of VT/SCD</td>
</tr>
<tr>
<td>Surgically repaired TOF</td>
<td>NSR; PVCs; IART 10%; VT 12%</td>
<td>Normal or mild ↑</td>
<td>Normal or RAD; LAD 5% to 10%</td>
<td>RBBB 90%</td>
<td>Possible P waves; RAH possible</td>
<td>RVH possible if RVOT obstruction or PHT</td>
<td>Absent sinus node in LAC; AV block with L-loop or AVCO</td>
</tr>
<tr>
<td>Congenitally corrected TGA</td>
<td>NSR</td>
<td>1° AVB &gt;50%; AVB 2%/year</td>
<td>LAD</td>
<td>Absence septal q; Q in III, aVF, and right precordium</td>
<td>Not if no associated defects</td>
<td>Not if no associated defects</td>
<td>Anterior AVN; positive T precordium; WPW with Ebstein’s</td>
</tr>
<tr>
<td>Complete TGA/intra-atrial baffle</td>
<td>Sinus brady 60%; EAR; junctional; IART 25%</td>
<td>Normal</td>
<td>RAD</td>
<td>Absence of q, small r, deep S in left precordium</td>
<td>Possible RAE</td>
<td>RSV; diminutive LV</td>
<td>Possible AV if VSD or TV surgery</td>
</tr>
<tr>
<td>UH with Fontan</td>
<td>Sinus brady 15%; EAR; junctional; IART &gt;50%</td>
<td>Normal in TA; 1° AVB in DILV</td>
<td>LAD in single RV, TA, single LV with noninverted outlet</td>
<td>Variable; ↑ R and S amplitudes in limb and precordial leads</td>
<td>RAH in TA</td>
<td>RSV with single RV; possible LWV with single LV</td>
<td>Absent sinus node in LAC; AV block with L-loop or AVCO</td>
</tr>
<tr>
<td>Dextrocardia</td>
<td>NSR; P-wave axis 105° to 165° with situs inversus</td>
<td>Normal</td>
<td>RAD</td>
<td>Inverse depolarization and repolarization</td>
<td>Not with situs inversus</td>
<td>LVH: tall R V₃–V₆; RSV: deep Q in V₁ and tail R right lateral</td>
<td>Sinus solitus: normal P-wave axis and severe CHD</td>
</tr>
<tr>
<td>ALCAPA</td>
<td>NSR</td>
<td>Normal</td>
<td>Possible LAD</td>
<td>Ant-leaflet Q waves; possible ant-sept Q waves</td>
<td>Possible LAE</td>
<td>Selective hypertrophy of posterobasal LV</td>
<td>Possible ischemia</td>
</tr>
</tbody>
</table>

AF indicates atrial fibrillation; ALCAPA, anomalous left coronary artery from the pulmonary artery; AVB, AV block; AVN, AV node; BHV, biventricular hypertrophy; CHD, congenital heart disease; DILV, double-inlet left ventricle; EAR, ectopic atrial rhythm; IART, intra-atrial reentrant tachycardia; LAD, left-axis deviation; LAE, left atrial enlargement; LAHB, left anterior hemiblock; LAI, left atrial isomerism; LV, left ventricle; LWV, left ventricular hypertrophy; NSR, normal sinus rhythm; PHT, pulmonary hypertension; PVC, premature ventricular contraction; RAD, right-axis deviation; RAE, right atrial enlargement; RBBB, right bundle-branch block (i., incomplete; c, complete); RV, right ventricle; RSV, right ventricular hypertrophy; RVOT, right ventricular outflow tract; RVP, right ventricular pressure; SCDO, sudden cardiac death; SVT, supraventricular tachycardia; TA, tricuspid atresia; TOF, tetralogy of Fallot; TV, tricuspid valve; UH, univentricular heart; VT, ventricular tachycardia; and WPW, Wolff-Parkinson-White syndrome.
small dimple lined with endocardium indicates the theoretical site of the absent tricuspid valve. The AV node is typically found on the floor of the right atrium bounded by the coronary sinus, tendon of Todaro, and right atrial dimple.\textsuperscript{11} It pierces the abnormally formed central fibrous body to become the His bundle along the left side of the septum.\textsuperscript{11} The His-Purkinje system’s remaining course is determined, in part, by the presence and location of associated VSDs. In general, the His bundle is further to the left and away from more anterior septal defects.\textsuperscript{11}

### Other Forms of Single-Ventricle Physiology

When the course of the conduction system and potential for AV block in other univentricular hearts are considered, key determinants include direction of ventricular looping and morphological type of dominant ventricle.\textsuperscript{12} The AV conduction system is commonly displaced in single ventricles with AV discordance and AVCD. In L-looped single left ventricles, 2 AV nodes may be present, with an inferior node that does not usually contact the ventricular septum.\textsuperscript{13,14} The elongated His bundle is susceptible to fibrous degeneration and complete AV block.\textsuperscript{13} With ventricular D-looping and a dominant right ventricle, the AV node is within Koch’s triangle, and the His bundle directly enters the ventricular septum.\textsuperscript{14}

### Electrocardiography in ACHD

#### Ostium Secundum Atrial Septal Defects

Atrial septum anomalies constitute >30% of ACHD.\textsuperscript{15} Typical ECG features are depicted in Figure 2. In unrepaired ostium secundum atrial septal defects (ASDs), the ECG rhythm is most commonly sinus, although atrial fibrillation and/or flutter increases with age and is found in ~20%.\textsuperscript{16,17} Surgical closure may reduce atrial arrhythmias, though less effectively in older patients.\textsuperscript{16–18} In a retrospective analysis, atrial arrhythmias persisted postoperatively in 60% of patients.\textsuperscript{16} All patients with persistent and new arrhythmias (2.3% at 3.8 years) were >40 years of age at surgery. In a study that randomized 521 patients who were ≥40 years of age to surgical closure versus medical therapy, no difference in new-onset atrial arrhythmias was noted at 7 years of follow-up.\textsuperscript{16} The impact of transcatheter ASD closure on atrial arrhythmias remains to be determined. In one retrospective study, all persistent arrhythmias remained after transcatheter device closure.\textsuperscript{18} The annual incidences of symptomatic paroxysmal and persistent atrial fibrillation or flutter were 17% and 11%, respectively, as predicted by older age (ie, ≥55 years of age).

An rSr\textsuperscript{19} or rsR\textsuperscript{19} QRS configuration is typically seen on the ECG in right precordial leads. This likely reflects right ventricular overload rather than true conduction delay.\textsuperscript{19} In

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**Figure 1.** Twelve-lead ECGs in a 37-year-old man with heterotaxy syndrome of the right atrial isomerism type. Note the difference in P-wave axis and morphology between A and B, indicating a shift in the governing pacemaker. A, The P-wave axis is 65°, positive in lead I, monophasic in inferior leads, and predominantly negative in V\textsubscript{5}. B, The P-wave axis is 105°, notched in inferior leads, and predominantly positive in V\textsubscript{1}.

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**Figure 2.** Fifteen-lead ECG in a 53-year-old woman with a secundum ASD. Note the rSr’ pattern over right precordial leads, with a broad and slurred r’. The QRS axis is vertical, and the PR interval is 200 ms. Encircled and magnified in the insert is the crochetage pattern in inferior leads, characterized by a notch near the R-wave apex.
contrast to a sharp and narrow \( r' \leq 5 \text{ mV} \) found in 2.4% of normal hearts, ASD is associated with a broader and somewhat slurred \( r' \). As a result of prolonged terminal forces, QRS duration is lengthened but often remains within the upper limit of normal. However, a complete right bundle-branch block pattern is not infrequent with increasing age. PR prolongation is seen in 6% to 19% of patients and uncommonly progresses toward higher-degree AV block,\(^{20}\) with complete block rarely reported.\(^{21}\) The QRS axis is usually between 0° and 180° and is often vertical (60° to 90°). Right-axis deviation is common in adults with associated pulmonary vascular disease.\(^{22}\) In contrast, left-axis deviation is rare but has been described in hereditary forms such as Holt-Oram syndrome and in older adults with presumably acquired left anterior fascicular block. Criteria for right atrial enlargement are fulfilled in 35% of patients.\(^{21}\)

In inferior leads, a notch near the apex of the R wave, coined “crochetage,” has been correlated with ASD.\(^{23}\) In 1560 adults and adolescents, 73.1% of patients with ASDs displayed this pattern, with a specificity of 92% when present in all inferior limb leads.\(^{24}\) Its incidence was higher in patients with larger defects and greater left-to-right shunting. Early disappearance was observed in 35.1% of surgically repaired patients, although the right bundle-branch block pattern persisted. In a second study, the crochetage pattern was observed in at least 1 inferior limb lead in 31.7% of preoperative patients with secundum ASDs.\(^{25}\) Specificity of the crochetage pattern was 86.1% and \( >92\% \) when present in at least 1 and in all 3 inferior leads, respectively.

### Ventricular Septal Defect

VSDs are the most commonly recognized congenital heart defects in children but often close spontaneously or cause congestive heart failure, prompting surgical closure before adulthood. In patients who have not undergone surgery, ECG findings more directly relate to hemodynamic consequences than to specific location.\(^{26}\) Newly diagnosed congenital VSDs in adulthood are most often small and restrictive or large and unrestrictive with Eisenmenger’s complex (ie, flow reversal in unrepaired complete AVCD).\(^{35}\) The most common presentations of AVCD in adulthood are partial defects, which consist of a primum ASD and cleft mitral valve, or surgically repaired complete AVCD. It is less common for a patient who has not undergone surgery with complete AVCD, including a large primum ASD, inlet VSD, and cleft mitral valve, to present with Eisenmenger’s physiology or with spontaneous closure of the inlet VSD by way of an aneurysm. In addition to manifestations from the displaced AV conduction system, ECG findings reflect the dominant physiology (eg, large left-to-right atrial shunt and mitral regurgitation in partial AVCD and Eisenmenger’s physiology in unrepaired complete AVCD).\(^{35}\) After surgical repair, atrial fibrillation or flutter may develop in \( \geq 5\% \) of patients.\(^{36}\) In the immediate postoperative period, persistent complete AV block occurs in 1% to 7% of patients and \( \geq 2\% \) of patients thereafter.\(^{36,37}\) Prolonged infra-Hisian conduction time may indicate increased risk of late AV block, even in the presence of a normal PR interval.\(^{38}\) Whereas increased ventricular ectopy has been described in up to 30% of patients, complex ventricular arrhythmias occur most commonly in the setting of left ventricular dysfunction.\(^{36}\)

Distinctive ECG features in the adult with AVCD (Figure 3) relate to the PR interval, right ventricular activation pattern, and QRS axis. First-degree AV block is found in \( >50\% \) of patients and is most commonly caused by intra-atrial conduction delay, although acquired postoperative AV block may occur.\(^{38}\) Electrophysiological studies reveal supra-Hisian first-degree AV block in 28% of patients and intra-atrial conduction delay in the majority.\(^{39}\) The QRS pattern in right precordial leads may resemble ostium secundum ASDs, with delay in right ventricular activation. Interestingly, this is thought to reflect conduction along a longer-than-normal right bundle branch that emanates from the inferiorly displaced His bundle, not from delayed parietal conduction.\(^{40}\) As
atrial fibrillation may occur in older individuals.42 Left atrial erately sized PDA, sinus rhythm is usually present, although opposed to moderate or large PDAs that may promote small in caliber and associated with a normal ECG, as deep S waves in V1 and tall R waves in leads V5 and V6, Left ventricular volume overload may be characterized by nonspecific repolarization changes.42,43

Patent Ductus Arteriosus
Most adults with moderate or large patent ductus arteriosus (PDA) will have undergone ductal ligation in infancy. Adults with unrepaired PDA generally fall into 1 of 2 categories: a fortuitously discovered PDA on echocardiography or Eisenmenger’s physiology with irreversible pulmonary vascular obstructive disease.35 Most incidentally identified PDAs are small in caliber and associated with a normal ECG, as opposed to moderate or large PDAs that may promote pulmonary hypertension and are accompanied by ECG findings dominated by Eisenmenger’s physiology. With a moderately sized PDA, sinus rhythm is usually present, although atrial fibrillation may occur in older individuals.42 Left atrial enlargement may be present, and PR prolongation is found in 10% to 20% of patients.43 The QRS axis is usually normal. Left ventricular volume overload may be characterized by deep S waves in V1, and tall R waves in leads V5 and V6, accompanied by nonspecific repolarization changes.42,43

Pulmonary Stenosis
Obstruction of the right ventricular outflow tract can occur at the pulmonary valve, below it, above it, in the main pulmonary artery trunk, or at 1 or both of its branches. These lesions represent a spectrum of unoperated and postoperative anomalies among the most frequently encountered in ACHD. Isolated congenital valvar pulmonary stenosis accounts for 10% of all congenital heart defects. In adults, pulmonary atresia, supravalvar, and branch pulmonary artery stenosis are common in the setting of tetralogy of Fallot. Congenital branch pulmonary artery stenosis can occur in isolation but is not usually associated with substantial right ventricular pressure overload. ECG features of primary infundibular stenosis or double-chambered right ventricle vary depending on the presence or absence of an associated VSD and/or concomitant valvar pulmonary stenosis.

Standard criteria for right ventricular hypertrophy are applicable to valvar pulmonary stenosis. In general, severity of pulmonary stenosis correlates with the R/S ratio in leads V1 and V6 and R-wave amplitude in lead V1.44 The PR interval is typically normal, but prolongation may reflect increased right atrial size and pressure.45 High-amplitude peaked P waves in lead II are commonly found in severe pulmonary stenosis, but not consistently so.46 The QRS axis may be normal with mild pulmonary stenosis but is deviated rightward with moderate or severe obstruction. Degree of right-axis deviation is positively correlated with right ventricular pressure.47

Aortic Coarctation
Complications in operated and unoperated adults with aortic coarctation include left ventricular hypertrophy, systemic hypertension, heart failure, aortic dissection, premature coronary artery disease, and cerebrovascular events.48 In a population-based study, only 1 of 536 survivors with aortic coarctation repair died suddenly during the first 20 years of follow-up.49 However, with longer-term follow-up, an additional 9 sudden deaths occurred, with all 7 presumably arrhythmic deaths occurring in patients with advanced ventricular dysfunction.49

In adults with uncomplicated aortic coarctation, the ECG usually exhibits normal sinus rhythm. Unlike children with concomitant left-to-right interatrial shunts, left but not right atrial enlargement is often seen.50 The PR interval is usually normal, and QRS axis is usually normal or displaced leftward. Persistent right ventricular hypertrophy beyond infancy is rare. Left ventricular hypertrophy is common, with the most sensitive ECG criterion being increased QRS voltage.51

Ebstein’s Anomaly
In Ebstein’s anomaly, the tricuspid valve is displaced api- cally, creating an atrialized portion that is morphologically and electrically right ventricle but functionally right atrium.52 Stimulation of this atrialized tissue may provoke ventricular arrhythmias, but spontaneous ventricular tachycardia is otherwise uncommon in the absence of associated malformations.53 Severity is determined in part by degree of tricuspid leaflet tethering, relative proportion of atrialized and true right ventricle, and presence or absence of right ventricular outflow tract obstruction. Patent foramen ovale and secur-
dum ASDs are common. In the adult, the ECG is helpful in establishing a new diagnosis, detecting accessory pathways, and characterizing arrhythmias, exemplified by Figure 4. Indeed, accessory AV or atriofascicular pathways are found in 25% of patients and are frequently right-sided and multiple.\textsuperscript{52} Supraventricular tachyarrhythmias, including AV reciprocating tachycardia, ectopic atrial tachycardia, and atrial fibrillation or flutter, occur in 30% to 40% of patients, constituting the most common presentation in adolescents and adults.\textsuperscript{54} Sudden death is reported in 3% to 4% of patients\textsuperscript{54} and may be preceded by cyanosis or rapid conduction of atrial fibrillation or flutter to the ventricles via high-risk or multiple pathways.\textsuperscript{55}

P waves are characteristically tall and broad (coined Himalayan) because of prolonged conduction in the enlarged right atrium.\textsuperscript{56} First-degree AV block often results from intra-atrial conduction delay, with a PR interval that can be markedly prolonged.\textsuperscript{57} Naturally, the PR interval may be shortened in the presence of an accessory AV or atriofascicular pathway. As the right ventricle is diminutive, low-amplitude QRS complexes are characteristically seen over right precordial leads.\textsuperscript{58} Because of the atrialized portion of the right ventricle, the QRS complex typically exhibits right ventricular conduction delay of the right bundle-branch type that is atypical and multiphasic.\textsuperscript{56} In unoperated patients, signal-averaged ECGs almost universally identify late potentials corresponding to delayed conduction across atrialized right ventricle.\textsuperscript{59} In the absence of ventricular preexcitation, the QRS axis is generally normal although occasionally leftward.\textsuperscript{60} Q waves are noted in lead V1 in about 50% of patients and may extend as far as lead V4.\textsuperscript{61,62} T-wave inversion in leads V1 to V4 is common.\textsuperscript{61,62} Deep Q waves may be present in leads II, III, and aVF.\textsuperscript{61} These Q waves are thought to reflect fibrotic thinning of the right ventricular free wall and/or septal fibrosis with coexisting left posterior hemiblock.\textsuperscript{61}

**Surgically Corrected Tetralogy of Fallot**

Tetralogy of Fallot is the most common cyanotic heart disease in adults.\textsuperscript{63} Corrective surgery involves atriotomy and/or ventricular incisions and patches, which predisposes patients to the late development of arrhythmias.\textsuperscript{64} Intra-atrial reentrant tachycardias are isthmus-dependent or incisional macroreentrant circuits, which may herald worsening ventricular function and tricuspid regurgitation.\textsuperscript{65,66} Moreover, sudden cardiac death is the most common cause of death late after repair.\textsuperscript{66} In a multicenter cohort study, 10% of patients developed atrial flutter, 11.9% of patients experienced sustained ventricular tachycardia (Figure 5), and 8.3% of patients died suddenly.\textsuperscript{64}

In adults, baseline sinus rhythm is the rule, with P waves of normal axis, duration, and amplitude, although somewhat peaked.\textsuperscript{67} The QRS axis is typically normal or displaced to
the right with right ventricular hypertrophy. In the presence of a right bundle-branch block, criteria for right ventricular hypertrophy include an R’ in V1 ≥15 mm and right-axis deviation of the initial vector representing unblocked forces.68 Left-axis deviation should raise suspicion for an associated AVCD, although left anterior hemiblock is present in 5% to 10% of adults.67 Right bundle-branch block is expected after repair, even in the absence of a ventriculotomy incision.67 Early QRS lengthening after repair results from surgical injury to the right bundle branch and myocardium,69 whereas later broadening reflects right ventricular dilation.70

Independent ECG predictors of ventricular tachycardia and sudden death include a QRS interval ≥180 ms (risk ratio 8.8) and annual increase in QRS duration (risk ratio 1.1 for each 1-ms increase per year).64 Patients with ventricular tachycardia or sudden death are more likely to have increased cardiothoracic ratios, at least moderate pulmonary and tricuspid regurgitation, and peripheral pulmonary stenosis. A greater degree of QT dispersion has also been noted, believed to reflect increased heterogeneity in myocardial repolarization.64 Other reported risk factors include frequent ectopic beats,71 increased right ventricular systolic pressures,72 complete heart block,73 and increased JT dispersion.74 Induction of sustained ventricular tachycardia is an imperfect but powerful predictor, independent of ECG and other clinical variables.75

Congenitally Corrected Transposition of the Great Arteries

Young adults with isolated congenitally corrected TGA may remain asymptomatic and undetected. The 12-lead ECG can provide the first clue to establishing this diagnosis (Figure 6). Given the displaced and fragile AV conduction system, complete AV block is estimated to occur in 2% of patients per year, irrespective of associated anomalies, with a prevalence of 22% on long-term follow-up.76 Electrophysiology studies reveal that the site of AV block is above or within the His bundle,77 a finding consistent with histopathological fibrosis of the His bundle.77 Clinically, a stable narrow QRS escape rhythm often accompanies complete AV block.76 Complete AV block follows surgical repair of an associated VSD in >25% of patients.76

Congenitally corrected TGA has been referred to as ventricular inversion, as the ventricles and associated bundle branches are reversed. The sinus node is normally positioned, generating a standard P-wave axis and morphology. However, the ventricular septum is depolarized in the opposite right-to-left direction, contributing to the pathognomonic ECG pattern.78 Septal Q waves are absent in left precordial leads and are found over right precordial leads. Similarly, anterior R waves are noted in left rather than right precordial leads. Moreover, as the septum is activated in a superior direction, broad Q waves are seen in leads III and aVF, most pronounced in lead III. Thus, left-axis deviation is the rule. Interestingly, positive T waves are present over all precordial leads in >80%, attributed to the side-by-side nature of the inverted ventricles.78 With associated Ebstein’s malformation of the left-sided tricuspid valve, left-sided accessory pathway may be present.79

Complete Transposition of the Great Arteries and Intra-Atrial Baffle

In complete TGA, the AV relationship is preserved with ventriculoarterial discordance. Complete TGA accounts for 5% to 7% of all congenital cardiac malformations.63 In 1959, Senning introduced an intra-atrial baffle repair without grafts or prostheses and, in 1964, Mustard proposed an alternative technique that used a pericardial patch.60 Although arterial switch surgery81 is now the procedure of choice, at the present time, most adults have intra-atrial baffles. Late arrhythmic complications include sinus node dysfunction, atrial tachyarrhythmias, and sudden cardiac death.49,82,83 Sinus node dysfunction and atrial flutter are noted in 60% and 24% of patients, respectively, at 20 years.82 Loss of coordinated atrial activity and rapid ventricular rates can result in hemodynamic compromise. Atrial arrhythmias are associated with impaired ventricular function84 and increased risk of sudden death in some but not all studies.85 Risk factors for sudden death include symptoms of arrhythmia or heart failure and documented atrial fibrillation or flutter.86 ECG criteria (other than atrial arrhythmias) have not been found to predict sudden death.

Characteristic ECG findings are illustrated in Figure 7.87 As sinus node dysfunction is highly prevalent, atrial and junctional rhythms may be seen. In simple complete TGA, AV node function is preserved. However, intra-atrial conduction delay may result in a prolonged PR interval. AV block is more common in the presence of a surgically repaired
tricuspid valve or associated VSD.88 Despite corrected physiology, the right ventricle remains systemically positioned. Consonantly, ECG criteria for right ventricular hypertrophy are usually present, with right-axis deviation.87 Right atrial enlargement may likewise be noted.87 Conversely, the subpulmonary left ventricle is diminutive with decreased terminal forces reflected in the absence of q waves, small r waves, and deep S waves over left precordial leads.

**Single-Ventricle Physiology With Fontan Surgery**

Developed in 1971 as surgical palliation for tricuspid atresia, the Fontan procedure has been adapted to various forms of single-ventricle physiology.12 Atrial arrhythmias may be associated with substantial morbidity and death, as hemodynamic deterioration can ensue. Depending on the type of repair, intra-atrial reentrant tachycardias or atrial fibrillation may occur in up to 57% of patients,89 with often complex and/or multiple circuits.90 At mid-term follow-up, sinus node dysfunction occurs in 13% to 16% of patients with classic modified Fontans (ie, right atrium to pulmonary artery anastomosis) and increases with duration of follow-up.91

Given the heterogeneity of single ventricles, the ECG appearance is highly variable, as illustrated in Figure 8. In patients with tricuspid atresia, the PR interval is usually normal, with tall and broad P waves. Left-axis deviation is characteristic.92 In the absence of a functional right ventricle, left ventricular forces are unopposed, as manifested by small r waves and deep S waves over the right precordial leads and tall R waves over the left precordial leads. In the most common subtype of double-inlet left ventricle (ie, with ventriculoarterial discordance), AV conduction is often abnormal, with PR prolongation and increased risk of complete heart block.93 As in congenitally corrected TGA, Q waves are absent over left precordial leads and may be present over right precordial leads. Q waves may also be seen in leads II, III, and aVF.93 In a series of patients with univentricular hearts of right ventricular morphology, all had right ventricular hypertrophy, and 61% of patients had a superior frontal QRS axis.94

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Figure 7. ECG examples in adults with complete TGA and Mustard baffles. A, Twelve-lead ECG in a 27-year-old man. The atrium is paced because of sinus node dysfunction. Note the right-axis deviation and right ventricular hypertrophy. The absence of a q wave, small r wave, and deep S wave in lead V6 reflect the diminutive left ventricle. B, Twelve-lead ECG in a 38-year-old woman. Note the intra-atrial reentrant tachycardia cycle length of 215 ms with 2:1 AV conduction.

Figure 8. ECG examples in adults with univentricular hearts. A, Twelve-lead ECG in a 31-year-old woman with tricuspid atresia, ASD, and VSD, status after old-style Fontan later revised to an extracardiac conduit. An intra-atrial reentrant tachycardia with a ventricular response rate of 128 bpm is seen. Note the characteristic left-axis deviation. B, Twelve-lead ECG in a 24-year-old man with mitral atresia and hypoplastic left ventricle status after classic Fontan. Note the slow intra-atrial reentrant tachycardia (cycle length 570 ms) with 2:1 AV conduction. The frontal QRS axis is superior with right ventricular hypertrophy.
Cardiac Malpositions

In dextrocardia with situs inversus (also known as mirror-image dextrocardia), the ventricles, atria, and viscera are inverted. The diagnosis is often fortuitous, as the heart usually functions normally. In mesocardia, the heart is centrally located in the chest with normal atrial and visceral anatomy. The apex is central or displaced to the right on chest radiography. Typically, no associated cardiac malformations are present. With dextrocardia and situs solitus (Figure 9), the ventricles are inverted but not the viscera or atria. Associated severe cardiac defects are common.

The ECG is diagnostically important in dextrocardia with situs inversus, as exemplified by Figure 10. The right atrium and sinus node are on the patient’s left side and yield a P-wave axis that remains inferiorly oriented but displaced to the right. In the absence of an ectopic focus, P waves are upright in aVR and inverted in I and aVL. Ventricular depolarization and repolarization occur in an inverse fashion. In lead I, the QRS is predominantly negative with T-wave inversion. Right precordial leads resemble left precordial leads of normal hearts, aVL resembles aVR, and vice versa. Thus, left ventricular hypertrophy is manifested by tall R waves in V1 and V2. Right ventricular hypertrophy is reflected by deeper Q waves and small R waves in lead I and taller R waves over right lateral chest leads (eg, V4R and V5R).

Figure 9. Twelve-lead ECG in a 45-year-old man with situs solitus, dextrocardia, anomalous pulmonary venous return, and pulmonary stenosis. The P-wave vector has an axis of 30°, consistent with atrial situs solitus and normal position of the sinus node. Note the reverse R-wave progression pattern, with decreasing amplitude from leads V1 to V6. The pathological Q wave in lead I may reflect right ventricular hypertrophy in the setting of pulmonary stenosis.

Figure 10. Fifteen-lead ECG in a 17-year-old man with dextrocardia, situs inversus, and a surgically repaired perimembranous VSD. The P-wave axis is directed rightward, at 135°. Note the inverse depolarization/repolarization pattern in limb leads with right axis deviation. An incomplete right bundle-branch block is noted with reverse R-wave progression from V4R to V7.
Coronary Anomalies

Isolated ectopic or anomalous origins of the coronary arteries are seen in 0.6% to 1.5% of patients undergoing coronary angiography.96 The prognosis is favorable if the anomalous coronary artery does not course between the pulmonary artery and aorta. Most common anomalies are not associated with myocardial ischemia and include ectopic origin of the left circumflex artery from the right sinus of Valsalva, anomalous origin of the right coronary artery from the left sinus, and anomalous origin of the left main coronary artery from the right sinus. In coronary–cameral fistulas, the ECG is typically normal until volume overload of the receiving chamber occurs with or without ischemic changes related to coronary steal.97

The ECG is useful in the diagnosis of anomalous origin of the left coronary artery from the pulmonary trunk. This is the most common congenital coronary anomaly associated with myocardial ischemia, with 25% of cases surviving to adolescence or adulthood.98 As a result of decreased perfusion pressure and hypoxemic blood flow through the left coronary artery, anterolateral myocardial infarction usually occurs before clinical recognition. The 12-lead ECG displays pathological Q waves in leads I, aVL, and V4 to V6, that are typically deep.99,100 Anteroseptal myocardial infarction patterns have also been described, as illustrated in Figure 11. In addition, the posterobasal portion of the left ventricle appears to selectively hypertrophy.100 This may result in left-axis deviation, a pattern consistent with left ventricular hypertrophy, and nonspecific repolarization changes.99,100

Conclusion

In conclusion, the ECG provides a wealth of information fundamental to clinical assessment in ACHD. In adults with previously undetected congenital heart disease such as ASD, Ebstein’s anomaly, congenitally corrected TGA, or cardiac malpositions, the ECG can provide important diagnostic clues. Serial ECGs may be of value in screening for hemodynamic ramifications of congenital malformations, particularly obstructive lesions. Moreover, in certain pathologies such as tetralogy of Fallot, individual measures and changing parameters provide clinically pertinent prognostic information for risk stratification. Finally, the ECG is vital in diagnosing and characterizing a broad spectrum of brady- and tachyarrhythmias that are frequently encountered in ACHD.

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References


ically staged Fontan procedure. Circulation. 1998(suppl);98:II352-II358.

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