Prevalence and spectrum of abnormal electrocardiograms in patients with an isolated congenital left ventricular aneurysm or diverticulum

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Aims

Congenital left ventricular aneurysm (LVA) and diverticulum (LVD) are rare cardiac anomalies and can be associated with ECG abnormalities and rhythm disturbances. We sought to investigate the prevalence and the spectrum of ECG abnormalities in such patients.

Methods and results

We assessed 125 patients with isolated LVA or LVD for the prevalence of ECG abnormalities and compared the findings to an age- and gender-matched control group. The 12-lead ECG patterns were evaluated according to commonly used criteria and were classified into three subgroups (distinct, mildly, and minor). Fifty-four of the 125 patients (43.2%) had normal and 71 (56.8%) abnormal ECGs. Mean age was 66 years. Forty-nine (39.2%) were male. Distinct abnormal ECG patterns were more prevalent in patients with LVD (38.2 vs. 15.8%, \( P = 0.04 \)), and apical location of the anomaly (36.6 vs. 16.6%, \( P = 0.02 \)). Older age (>66 years) was associated with a trend for a higher prevalence of abnormal ECG pattern (33 vs. 18%, \( P = 0.06 \)), whereas gender had no influence (32 vs. 16%, \( P = 0.14 \)). This study also shows that the sensitivity, specificity, positive predictive value and negative predictive value of a 12-lead ECG for the diagnosis of LVA or LVD are low.

Conclusion

This large single-centre study suggests that the prevalence of abnormal ECG patterns in patients with isolated LVA or LVD is as high as 56.8%. However, ECG is not specific and sensitive to be used as a screening tool in such patients.

Keywords

Left ventricle • Aneurysm • Diverticulum • Abnormal ECG • Congenital • Prevalence

Introduction

Congenital left ventricular aneurysm (LVA) and diverticulum (LVD) are rare malformations with ~500 cases in the literature since the first description in 1816. A recent publication demonstrated a prevalence of 0.76% in patients undergoing coronary angiography. Although most investigators distinguish between LVA and LVD, the cause, histopathology, and clinical implications remain controversial. The advent of echocardiography and magnetic resonance imaging has led to earlier diagnosis, including prenatal detection. The published research on these anomalies consists, beside one large single-centre study, of case reports and small case series, including mostly infants and young children with large aneurysms and poor clinical outcome. High morbidity and mortality rates due to heart failure, aneurysm rupture, thrombo-embolism, and sudden death of unclear cause have been reported. More recently, published data demonstrated a more favourable course with no cardiac mortality in adults with congenital LVA and LVD. A significant proportion of the published literature report arrhythmias in patients with these anomalies, ranging from frequent premature ventricular complexes to sustained supraventricular or ventricular tachycardia. Recently, a large single-centre study demonstrated a significantly higher frequency of rhythm disturbances in patients with congenital LVA/LVD compared with control.© The Author 2009. For permissions please email: journals.permissions@oxfordjournals.org
The purpose of this study was two-fold. On the one hand, we assessed the prevalence and the spectrum of ECG abnormalities detected in a large group of patients with isolated congenital LVA or LVD. On the other hand, we investigated whether a single ECG abnormality or combinations of two ECG criteria allow the diagnosis of congenital LVA/LVD.

Methods

Definition

The diagnosis of congenital LVA or LVD in our series was established after angiographic exclusion of coronary artery disease, and clinical exclusion of local or systemic inflammation or traumatic causes as well as cardiomyopathies. Definition of an LVA included a- or dyskinetic structures with a ratio of the connection to the left ventricular cavity compared with the maximum diameter of the body of the anomaly being >1. In LVD, this ratio is <1 in a structure with normal systolic contraction.

Study population

Between 1 January 2001 and 31 December 2003, 17,257 consecutive patients underwent diagnostic coronary angiography at our institution. During this time, 125 patients with angiographically proven isolated congenital LVA or LVD were identified. Their mean age was 66 years (22–94). Forty-nine (39.2%) were male. Fifty-seven patients were found to have congenital LVAs, and in 68 patients, congenital LVDs were diagnosed. Patients found to have LVA or LVD presented on clinical evaluation with a wide range of symptoms. The most common complaints were typical angina or atypical chest pain in 57% of all cases. The second most frequent cause for invasive evaluation was syncope, rhythm disturbances, or evaluation for valvular disease. The criteria for such a designation included 25–28 by two independent cardiologists. We arbitrarily classified ECGs into three subgroups as suggested by Pelliccia et al. on the basis of the presence of more than one of the listed criteria. The subgroups were as follows (Figures 1–3).

Distinctly abnormal electrocardiogram

Distinctly abnormal ECGs were those that were strongly suggestive of cardiovascular disease. The criteria for such a designation included the following: (i) striking increase in R- or S-wave voltage (>35 mm) in any lead; (ii) Q-waves >4 mm in depth and present in more than two leads; (iii) repolarization pattern with inverted T-wave >2 mm in more than two leads; (iv) left bundle branch block; (v) marked left (<−30°) or right (>110°) QRS axis deviation; and (vi) Wolf–Parkinson–White’s or Brugada’s pattern, and e-wave.

Mildly abnormal electrocardiogram

Mildly abnormal ECGs were those compatible with the presence of cardiovascular disease. The criteria for such a designation included the following: (i) increased R- or S-wave voltage (30–34 mm); (ii) Q-waves 2–3 mm in depth and present in more than two leads; (iii) repolarization patterns with either flat, minimally inverted, or particularly tall (i.e. >15 mm) T-waves in more than two leads; (iv) abnormal R-progression in the anterior precordial leads; (v) right bundle branch block (QRS pattern, QRS >120 ms in V1 and V2); (vi) right atrial enlargement (pulled P-waves >2.5 mm in leads II, III, or V1); (vii) left atrial enlargement (pulled positive P-wave in lead II and/or deep prolonged negative P-wave in V1); and (viii) short PR interval (<120 ms), atrial fibrillation.

Normal electrocardiogram or electrocardiogram with minor alterations

This subgroup consisted of ECGs that were completely normal and those with minor alterations that has been reported, e.g. in trained athletes. These minor alterations included the following: (i) increased PR interval duration (>0.20 s); (ii) mild increase in R- or S-wave voltage (25–29 mm); (iii) early repolarization (ST-elevation >2 mm in more than two leads); (iv) incomplete right bundle branch block (RSR pattern in V1 and V2 of <120 ms in duration); and (v) sinus bradycardia (<60 bpm).

Control group

The control group was established by collecting data of 125 age- and gender-matched individuals without the diagnosis of isolated congenital LVA or LVD and without evidence for coronary artery disease, previous non-coronary cardiac surgery, local or systemic inflammation, or cardiomyopathies. The patients underwent coronary angiography for various reasons at our institution between 1 January 2001 and 31 December 2003. Prevalence of obesity was 27%, and 23% were smokers. Hypertension was present in 38% and diabetes in 7% of the control group.

Statistical analysis

Variables were reported as mean ± SD for continuous variables or percentages. Normal distribution of variables was assessed with the Kolmogorov–Smirnov test. Comparisons for continuous variables were performed using the t-test and by using the χ² test or Fisher exact test for categoric variables, as appropriate. A value of P < 0.05 was considered to be statistically significant. Sensitivity, specificity, positive predictive value, and negative predictive value were calculated for every single distinct ECG abnormality and combinations of two distinct ECG abnormalities to assess the utility of the ECG for screening for LVA or LVD.

Results

Prevalence and pattern of electrocardiographic abnormalities

Abnormal ECGs were identified in 71 patients (56.8%); these included 35 (28%) with distinctly abnormal and 36 (28.8%) with mildly abnormal pattern. Of the remaining 54 patients (43.2%), ECGs were completely normal in 14 patients (11.2%) or showed only minor alterations in 40 (32%). In patients with the diagnosis of isolated congenital LVA or LVD, T-wave alterations in the precordial leads, Q-waves (2–3 mm), abnormal R-progression in the anterior precordial leads, atrial fibrillation, complete and incomplete right bundle branch block, early repolarization pattern, and PR interval >200 ms were more prevalent compared with control. The prevalence of various abnormalities in the three ECG subgroups is summarized in Table 1. The values of various conduction intervals [PQ interval, QRS complex, QT interval,
Electrocardiograms in general were not able to establish a decisive and accurate diagnosis of congenital LVA or LVD. The combination of two distinct abnormal ECG abnormalities with the highest positive predictive value (negative T-waves in precordial leads plus left or right axis deviation) showed a sensitivity of 0.16, a specificity of 0.07, a positive predictive value of 0.2, and a negative predictive value of 0.08, respectively. The combination of two distinct abnormal ECG abnormalities with the highest negative predictive value (left bundle branch block plus Q-wave >4 mm) showed a sensitivity of 0.1, a specificity of 0.6, a positive predictive value of 0.3, and a negative predictive value of 0.51, respectively.

**Discussion**

Several reports over the past decades have described a wide range of rhythm disturbances and a variety of ECG alterations in patients with congenital LVA/LVD. In the present study, we assessed the prevalence of ECG abnormalities in the largest series of patients with isolated congenital LVA or LVD to date. Furthermore, we investigated whether a single ECG abnormality or a combination of two ECG criteria allows the diagnosis of congenital LVA/LVD.

Our investigation shows that distinct or mild ECG abnormalities, raising clinical suspicion for underlying cardiac disease, were present in 56.8% of these patients. This proportion is greater than previously reported in a population without congenital LVA or LVD and without evidence of coronary artery disease aged 45–75 years. The distinct or mild ECG abnormalities included mainly T-wave inversion, abnormal Q-waves, altered R-progression in the precordial leads, atrial fibrillation, or marked left (≤−30°) or right (≥110°) axis deviation. The remaining ECGs with minor abnormalities included early repolarization.

**Table 1** Prevalence and spectrum of electrocardiographic abnormalities in patients with congenital left ventricular aneurysm or diverticulum, and controls

<table>
<thead>
<tr>
<th>Parameter</th>
<th>LVA/LVD group, n (%)</th>
<th>Control group, n (%)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Distinctly abnormal</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>R- or S-wave &gt;35 mm</td>
<td>1 (0.8)</td>
<td>5 (4.0)</td>
<td>0.1</td>
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<tr>
<td>Negative T-waves in precordial leads</td>
<td>17 (13.4)</td>
<td>3 (2.4)</td>
<td>0.003</td>
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<tr>
<td>Q-wave: &gt;4 mm</td>
<td>8 (6.4)</td>
<td>3 (2.4)</td>
<td>0.14</td>
</tr>
<tr>
<td>Left or right axis deviation</td>
<td>11 (8.8)</td>
<td>6 (4.8)</td>
<td>0.24</td>
</tr>
<tr>
<td>Pre-excitation pattern, e-wave, Brugada’s pattern</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1</td>
</tr>
<tr>
<td>Left bundle branch block</td>
<td>4 (3.2)</td>
<td>2 (1.6)</td>
<td>0.4</td>
</tr>
<tr>
<td>Mildly abnormal</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>R- or S-wave 30–34 mm</td>
<td>2 (1.6)</td>
<td>3 (2.4)</td>
<td>0.7</td>
</tr>
<tr>
<td>Flat/tall T-wave</td>
<td>34 (27.2)</td>
<td>4 (3.2)</td>
<td>0.001</td>
</tr>
<tr>
<td>Q-wave 2–3 mm</td>
<td>26 (20.8)</td>
<td>5 (4.0)</td>
<td>0.001</td>
</tr>
<tr>
<td>Left atrial enlargement</td>
<td>5 (4.0)</td>
<td>5 (4.0)</td>
<td>1</td>
</tr>
<tr>
<td>Right atrial enlargement</td>
<td>4 (3.2)</td>
<td>1 (0.8)</td>
<td>0.2</td>
</tr>
<tr>
<td>Abnormal</td>
<td>41 (32.8)</td>
<td>15 (12.0)</td>
<td>0.002</td>
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<td>R-progression in the anterior precordial leads</td>
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<tr>
<td>PR interval &lt;120 ms</td>
<td>2 (1.6)</td>
<td>2 (1.6)</td>
<td>1</td>
</tr>
<tr>
<td>Atrial fibrillation</td>
<td>25 (20)</td>
<td>3 (2.4)</td>
<td>0.001</td>
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<tr>
<td>Right bundle branch block</td>
<td>8 (6.4)</td>
<td>1 (0.8)</td>
<td>0.02</td>
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<tr>
<td>Minor alterations</td>
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<tr>
<td>R- or S-wave 25–29 mm</td>
<td>8 (6.4)</td>
<td>6 (4.8)</td>
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<td>Early repolarization</td>
<td>33 (26.4)</td>
<td>6 (4.8)</td>
<td>0.001</td>
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<tr>
<td>Incomplete right bundle branch block</td>
<td>20 (16)</td>
<td>4 (3.2)</td>
<td>0.002</td>
</tr>
<tr>
<td>PR interval &gt;200 ms</td>
<td>12 (9.6)</td>
<td>4 (3.2)</td>
<td>0.05</td>
</tr>
<tr>
<td>Sinus bradycardia</td>
<td>20 (16)</td>
<td>13 (10.4)</td>
<td>0.3</td>
</tr>
</tbody>
</table>

Bold values denote statistically significant results.

Electrocardiographic patterns did not differ with respect to gender, although a larger proportion of female patients had distinctly abnormal ECG compared with male patients (32 vs. 16%, P = 0.14). Conversely, a larger proportion of male patients showed normal ECGs (53 vs. 36%, P = 0.27). Patients at older age (>66 years) were more likely to have distinct abnormal ECG pattern (33 vs. 18%), although this did not reach statistical significance (P = 0.06).

**Electrocardiographic patterns in relation to type of abnormality**

Electrocardiographic patterns also differed with respect to the type of abnormality (Figure 4). Distinctly abnormal ECGs were more frequently encountered in patients with LVD (38 vs. 16%, P = 0.04), whereas mildly abnormal and normal ECG patterns were equally distributed [49.1% (LVA) vs. 34% (LVD), P = 0.44].

**Electrocardiographic patterns in relation to location of abnormality**

Electrocardiographic patterns differed also with respect to the location of the anomaly (Figure 5). We arbitrarily classified the location of the anomalies into four subgroups [anterior (n = 16), inferior (n = 32), apical (n = 71), and other (n = 6)]. A larger proportion of distinct abnormal ECG was encountered in the group with apical location compared with non-apical location of the anomaly (37 vs. 17%, P = 0.02).
Figure 1  Example of a distinctly abnormal electrocardiogram (repolarization pattern with inverted T-wave >2 mm in more than two leads).

Figure 2  Example of a mildly abnormal electrocardiogram [increased R- or S-wave voltage (30–34 mm)].
pattern, incomplete right bundle branch block, and sinus bradycardia, which are commonly regarded as innocent electrocardiographic changes, e.g., associated with trained individuals. The prevalence of abnormal ECGs in our series was larger in female compared with male individuals without reaching statistical significance. This is in contrast to findings in a large unselected study with more than 4800 individuals without coronary artery disease, reporting a larger proportion of abnormal ECGs in male individuals. Correspondingly, studies of Pelliccia et al. also demonstrated a higher prevalence of abnormal ECGs in trained male athletes. Data specifically addressing this topic in patients with congenital LVA or LVD are lacking.

In this analysis, we could demonstrate a trend ($P = 0.06$) for the influence of age on the prevalence of distinct or mild ECG abnormalities. In younger patients (<66 years), the largest proportion of the ECG anomalies includes early repolarization pattern, incomplete right bundle branch block, and sinus bradycardia, whereas in the older patients, abnormal repolarization patterns.

**Figure 3** Example of an electrocardiogram with minor alterations (early repolarization pattern).

**Figure 4** Distribution of three electrocardiographic categories with respect to the type of abnormality among 125 patients with congenital left ventricular aneurysm or diverticulum.

**Figure 5** Distribution of three electrocardiographic categories with respect to the location of the abnormality among 125 patients with congenital left ventricular aneurysm or diverticulum.
(e.g. inverted T-waves), abnormal Q-waves (>4 mm), and marked left (<−30°) or right (>110°) axis deviation are prevalent in 16.8%. However, an increasing prevalence of ECG anomalies is commonly associated with advanced age, also reported in individuals without congenital LVA or LVD reaching ~10% in the age group 65–75 years.29,31

We also demonstrated a higher prevalence of ECG abnormalities in patients with congenital LVD compared with LVA (P = 0.04). This is consistent with a recently published series of patients with congenital LVA and LVD, where the incidence of rhythm disturbances during a 5-year follow-up period was significantly higher in the congenital LVD group compared with the congenital LVA group.2 However, the causal connection between a localized diverticulum and ECG changes remains unclear, and the assumption of any associated cardiac remodelling process in addition to the presence of LVD remains speculative. The majority of statistically significant differences to the control group in the prevalence of ECG abnormalities included alterations of parameters of de- and repolarization (PR interval, abnormal R-progression and T-wave abnormalities in the anterior precordial leads, incomplete and complete right bundle branch block, and early repolarization pattern). As congenital LVAs and LVDs are thought to be developmental anomalies, starting on the fourth embryonic week and can be explained by a partial stop in the development of the embryonic ventricle.32 Before this, a development disturbance of the primitive paramidline mesoderm occurs between the 14th and 18th day of the embryonic phase, shortly after the differentiation into a ventral and parietal part.33,34 Considering this, one could hypothesize that the development of the intracardiac conduction system might be affected as well during this phase of embryonic development. Finally, in patients with the diagnosis of congenital LVA or LVD, cardiac dextroversion is described in ~10% and might explain the frequent occurrence of axis deviation in our series. However, the basic mechanism responsible for the distinct ECG abnormalities in patients with congenital LVA or LVD remains unresolved, and multiple causes are most likely.

The prevalence of distinct abnormal ECG changes was significantly higher in patients with an apical location compared with non-apical location of the anomaly (P = 0.02). Data from the literature in this respect are lacking, but >80% of the anomalies located at the left ventricular apex in our series were LVDS, which are themselves associated with a higher prevalence of distinct abnormal ECGs. Therefore, it is not surprising that apical location of the anomaly has a higher prevalence of abnormal ECGs. This study shows that the sensitivity, specificity, positive predictive value, and negative predictive value of a complete 12-lead electrocardiogram for the diagnosis of congenital LVA or LVD are low. Neither one single distinct ECG abnormality nor any combination of two distinct ECG abnormalities allows a decisive and accurate diagnosis making the ECG an inadequate screening tool for such patients.

**Study limitation**

Several limitations of the study merit further discussion. First, this study is subject to limitations inherent in retrospective studies. Secondly, we do not have any information regarding dynamic changes in the ECG, as only one 12-lead ECG tracing was available per patient. So, we cannot report whether these ECG abnormalities worsen over time or they might disappear.

**Conflict of interest:** none declared.

**References**


