DOUBLE-CHAMBERED LEFT VENTRICLE: CLINICAL FEATURES COMPARISON BETWEEN CHILDREN AND ADULTS

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ABSTRACT

Double-chambered left ventricle is a rare congenital heart defect. The clinical features, diagnosis, treatment and patient prognosis of this lesion have not been sufficiently elaborated. The present systematic review found that the accessory ventricular septum was often an abnormal muscle band, and sometimes a membranous structure, a fibromuscular ridge, or prominent trabeculations. Less than one-third of the patients were associated with other congenital heart defects. Diagnosis of double-chambered left ventricle can usually be made by transthoracic echocardiography; however, it might be misdiagnosed as atrial or ventricular septal defect. The accessory chamber was often smaller than the main chamber, and often had wall thinning. The accessory chamber wall dysfunction/hypokinesis was seen in half of the cases. One-third patients had left ventricular outflow tract obstruction. The adult patients showed abnormal electrocardiographic findings more than in pediatrics. Nevertheless, no differences were found between adult and pediatric patients in terms of left ventricular function and structure, treatment of choice, and patient outcomes. Most patients were asymptomatic with no left ventricular obstruction, and thus do not need surgical treatment. Surgical resection of the accessory ventricular septum is warranted when the patients become symptomatic as a result of left ventricular obstruction, or associated with other congenital heart defects. The patients’ outcomes are promising. Differential diagnosis should be made from other types of left ventricular outpouching, other congenital heart defects, and left ventricular non-compaction.

Key Words: Diagnosis, Differential, Heart defects, Congenital, Ventricular outflow obstruction.

RESULTS

The 29 case reports, which dealt with a single patient in each, included in total 29 patients. One case was a 24-week gestation fetus, whose gender was not reported. For the remaining 28 patients, 20 (71.4%) were males and 8 (28.6%) were females with a male-to-female ratio of 2.5:1. Their mean age was 28.6 ± 11.7 years (n=28). There was no age difference between the male and the female patients (28.2 ± 18.0 years vs. 25.4 ± 17.0, p=0.758).

There were 10 (34.5%) pediatric patients (including the fetal case), and unspecified (n=1). The size of heart defects, and 19 (65.5%) patients were available: the ECG findings were normal in 6 (30%) patients.

In 20 patients, their electrocardiographic (ECG) findings were normal in 6 (30%) patients. In the 6 patients with a normal ECG, 3 (50%) were pediatric and 3 (50%) were adults (χ²=0.00, p=1.000). There were 31 clinical presentations in 15 symptomatic patients, where the most common symptoms were chest pain and palpitation. Their duration of symptoms was 22.5 ± 25.1 months (n=5). Heart murmur was described in 13 (13/29, 44.8%) patients: in 4 (4/13, 30.8%) patients, no heart murmur was audible, and in 9 (69.2%) patients, a systolic murmur was heard (χ²=3.85, p=0.05). The locations of the murmurs were: left sternal border (n=4), apex (n=2), base (n=1), and unspecified (n=1).

Among 14 asymptomatic patients, 6 (42.9%) were pediatrics and 8 (57.1%) were adults (χ²=0.57, p=0.706). There were 31 clinical presentations in 15 symptomatic patients, where the most common symptoms were chest pain and palpitation. Their duration of symptoms was 22.5 ± 25.1 months (n=5). Heart murmur was described in 13 (13/29, 44.8%) patients: in 4 (4/13, 30.8%) patients, no heart murmur was audible, and in 9 (69.2%) patients, a systolic murmur was heard (χ²=3.85, p=0.05). The locations of the murmurs were: left sternal border (n=4), apex (n=2), base (n=1), and unspecified (n=1).

In 20 patients, their electrocardiographic (ECG) findings were available: the ECG findings were normal in 6 (30%) patients. In the 6 patients with a normal ECG, 3 (50%) were pediatric and 3 (50%) were adults (χ²=0.00, p=1.000); while 3 (21.4%) patients with an abnormal ECG were pediatric and 3 (50%) were adult cases (χ²=3.85, p=0.05). In the 6 patients with a normal ECG, 3 (50%) were pediatric and 3 (50%) were adults (χ²=0.00, p=1.000); while 3 (21.4%) patients with an abnormal ECG were pediatric and 3 (50%) were adult cases (χ²=3.85, p=0.05).

Fourteen patients (48.3%) were asymptomatic, and unspecified (n=1). The size of heart defects, and 19 (65.5%) patients were available: the ECG findings were normal in 6 (30%) patients. In 20 patients, their electrocardiographic (ECG) findings were normal in 6 (30%) patients. In the 6 patients with a normal ECG, 3 (50%) were pediatric and 3 (50%) were adults (χ²=0.00, p=1.000). There were 31 clinical presentations in 15 symptomatic patients, where the most common symptoms were chest pain and palpitation. Their duration of symptoms was 22.5 ± 25.1 months (n=5). Heart murmur was described in 13 (13/29, 44.8%) patients: in 4 (4/13, 30.8%) patients, no heart murmur was audible, and in 9 (69.2%) patients, a systolic murmur was heard (χ²=3.85, p=0.05). The locations of the murmurs were: left sternal border (n=4), apex (n=2), base (n=1), and unspecified (n=1).

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Table I: The associated congenital heart defects and secondary cardiac anomalies.

<table>
<thead>
<tr>
<th>Associated Cardiac Defect</th>
<th>n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventricular septal defect</td>
<td>15 (51.7%)</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>10 (34.5%)</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>9 (31.0%)</td>
</tr>
<tr>
<td>Hypertension</td>
<td>7 (24.1%)</td>
</tr>
<tr>
<td>Coronary artery disease</td>
<td>4 (13.8%)</td>
</tr>
<tr>
<td>Pulmonary hypertension</td>
<td>3 (10.3%)</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>3 (10.3%)</td>
</tr>
<tr>
<td>LVOTO</td>
<td>2 (6.9%)</td>
</tr>
<tr>
<td>Mitral stenosis</td>
<td>2 (6.9%)</td>
</tr>
<tr>
<td>Atrial fibrillation</td>
<td>1 (3.4%)</td>
</tr>
</tbody>
</table>

Table II: The nature of the accessory septum of the left ventricle.

<table>
<thead>
<tr>
<th>Accessory septum</th>
<th>n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>A muscle band</td>
<td>6 (56.3%)</td>
</tr>
<tr>
<td>A membranous structure</td>
<td>4 (36.4%)</td>
</tr>
<tr>
<td>Prominent trabeculations</td>
<td>3 (27.3%)</td>
</tr>
</tbody>
</table>

Besides, 2 (2/29, 6.9%) patients had genetic problems: MYH7 rare variant, and VACTERL association.

The dimensions of the accessory chamber of the left ventricle was (described as long axis × short axis, mm²) measured as 1898 ± 324.5 mm² vs. 1925.2 ± 55.2 mm², (p=0.011). An adult patient was reported to have an end diastolic volume of the accessory left ventricle of 36 ml/m². 

The pressure gradient decreased to 14.3 ± 16.9 mmHg (n=3) after treatment.
Double-chambered left ventricle

Table III: A comparison between left ventricular aneurysm and diverticulum.

<table>
<thead>
<tr>
<th>Feature</th>
<th>Aneurysm</th>
<th>Diverticulum</th>
<th>DCLV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Communicating opening&lt;sup&gt;4-15&lt;/sup&gt;</td>
<td>Wide neck</td>
<td>Narrow neck</td>
<td>By the anomalous septum or muscle bundle</td>
</tr>
<tr>
<td>Wall motion&lt;sup&gt;4,15&lt;/sup&gt;</td>
<td>Dyskinetic</td>
<td>Absence of dyskinetic motion</td>
<td>Normal or dyskinetic</td>
</tr>
<tr>
<td>Wall structure&lt;sup&gt;4,15&lt;/sup&gt;</td>
<td>Lack of full layers</td>
<td>Full layers</td>
<td>Full layers</td>
</tr>
<tr>
<td>Abnormal muscular and/or fiber bundles to division&lt;sup&gt;29&lt;/sup&gt;</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Associated congenital mid-line defects&lt;sup&gt;22&lt;/sup&gt;</td>
<td>Seldom</td>
<td>Often</td>
<td>Seldom</td>
</tr>
</tbody>
</table>

DCLV: Double-chambered left ventricle.

The nature of the accessory septum of the left ventricle was described for 20 patients, most of which were an abnormal muscle band (Table II). The accessory septum could be a complete,<sup>19</sup> or an incomplete structure,<sup>20,28</sup> Their course was horizontal in several cases,<sup>1,19</sup> and was Y-shaped in a patient.<sup>16</sup>

Other diagnostic techniques to confirm the diagnosis of DCLV included magnetic resonance imaging (n=19), catheterisation/coronary angiography (n=8), 3-D echocardiography (n=3), and contrast echocardiography (n=1).

One patient died of accident before diagnosis of DCLV was made.<sup>14</sup> Besides, management of 14 (14/29, 48.3%) patients was mentioned: 11 (78.6%) were conservatively and 3 (21.4%) were surgically treated (χ²=9.14, p=0.007).

The surgical indications of the 3 patients undergoing a surgical treatment were an intra-left ventricular pressure gradient of 110 mmHg,<sup>11</sup> a high flow velocity of >2 m/s,<sup>1</sup> and severe left ventricular outflow obstruction with necessity of concurrent Ross procedure.<sup>19</sup> The conservative treatment of choice was based on that patient were asymptomatic and without left ventricular outflow obstruction.

The outcomes of 7 (50%) patients were reported: 6 (85.7%) were recovered and 1 (14.3%) was improved. There were no differences between conservative and surgical treatments (χ²=0.29, p=1.000) and between pediatric and adult patients (χ²=0.19, p=1.000) in terms of pateints’ outcomes (namely, event-free survival).

**DISCUSSION**

DCLV is a very rare congenital heart defect, and it is characterised by a division of the ventricular cavity by an anomalous septum of DCLV or muscle bundle.<sup>17</sup> The accessory chamber of the left ventricle may have a thinned ventricular wall and hypokinesis. The impaired wall contractility serves to distinguish the DCLV from left ventricular aneurysm.

The embryology of DCLV remains uncertain. But it was proposed that DCLV might be the result of incomplete regression of the trabeculations, probably a variant of left ventricular non-compaction.<sup>15,32</sup>

Differential diagnosis of DCLV includes diverticulum and aneurysm of the left ventricle. Both ventricular aneurysm and ventricular diverticulum do not have the abnormal muscular and (or) fiber bundles for left ventricular division.<sup>29</sup> In left ventricular aneurysm, as a result of myocardial infarction, the aneurysmal wall shows delayed gadolinium enhancement with impaired wall motion, either akinetic or dyskinetic. Left ventricular diverticulum contains all three layers of the myocardial tissues, and the connection to the left ventricular cavity is narrow. DCLV is also composed of all three layers of the myocardial tissues but there is a muscular or membranous accessory septum to separate the ventricle. The differential diagnoses between the three forms of left ventricular outpouching are of clinical importance as for the different management policies and patient outcomes (Table III). Left ventricular aneurysm and diverticulum may sometimes lead to systemic embolisation, cardiac dysfunction, valve insufficiency, myocardial rupture, severe ventricular arrhythmias, and even cardiac death.<sup>32</sup> Moreover, the diagnosis of left ventricular non-compaction in which a giant trabecula, parallel to the interventricular septum, subdivided the left ventricle, thus simulating an accessory ventricular chamber.<sup>32</sup>

DCLV is usually asymptomatic and has a benign prognosis. Nevertheless, one case with coronary embolism and two cases with non-sustained ventricular tachycardia-ventricular fibrillation have been described.<sup>15</sup> When symptomatic or when associated with other cardiac abnormalities, surgical treatment is recommended.<sup>33</sup>

The present study revealed that the accessory ventricular septum was often an abnormal muscle band, and sometimes a membranous structure, a fibromuscular ridge, or prominent trabeculations. Less than one-third of the patients are associated with other congenital heart defects. Diagnosis of DCLV can usually be made by transthoracic echocardiography; however, it might be misdiagnosed as atrial or ventricular septal defect. The accessory chamber was often smaller than main chamber, and often has wall thinning. The accessory chamber wall dysfunction/hypokinesis was seen in half of the case. One-third patients had left ventricular outflow tract obstruction. The adult patients showed abnormal ECG findings more than in pediatrics. Nevertheless, no differences were found between adult and pediatric patients in terms of left ventricular function and structure, treatment of choice, and patient outcomes.

**CONCLUSION**

DCLV is a rare congenital heart defect. Most patients are asymptomatic with no left ventricular obstruction, and
thus do not need surgical treatment. Surgical resection of the accessory ventricular septum is warranted when the patients become symptomatic as a result of left ventricular obstruction, or associated with other congenital heart defects. The patients’ outcomes are promising. Differential diagnosis should be made from other types of left ventricular outpouching, other congenital heart defects, and left ventricular non-compaction.

CONFLICT OF INTEREST:
Author declared no conflict of interest.

AUTHOR’S CONTRIBUTION:
SMY: Substantial contribution to the conception and design of the work; and the acquisition, analysis, and interpretation of data for the work; drafting the work and revising it critically for important intellectual content; final approval of the version to be published; agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy and integrity of any part of the work are appropriately investigated and resolved.

REFERENCES

