SYSTEMATIC REVIEW ARTICLE

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 are 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 has 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 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.

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

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INTRODUCTION

Double-chambered left ventricle (DCLV) is a rare condition in which the left ventricular cavity is divided into two parts by an abnormal accessaory ventricular septum.[1] Due to its rarity, it may be misdiagnosed as simple congenital heart defects,[2] or confused with double-chambered right ventricle,[3] or with other types of left ventricluar outpouching.[4] Thus, this congenital heart defect has shown important clinical implications. Moreover, the clinical features of DCLV have not been sufficiently described. In order to give an overview of this rare lesion, a comprehensive review is made.

METHODOLOGY

The preferred reporting items for systematic reviews and meta-analyses (PRISMA) statement guidelines were followed in this meta-analysis. Publications were systematically searched in the PubMed, Highwire Press, and the Cochrane Library databases from January 2000 to December 2018. The MeSH terms and keywords used to identify articles included double-chambered, two-chambered, left ventricle, double-chambered left ventricle, anomalous muscle bundles, and accessory ventricular septum. The screening of the bibliographic references helped in completing the literature retrieval. Thirty-three articles related to the topic were found and keywords in the literature search; and 29 articles, which met the inclusion and exclusion criteria during preliminary assessment, were included in the review. The exclusion criteria were articles of: double-chambered right ventricle (n=2), patient was misdiagnosed as DCLV (n=1), and other types of left ventricular outpouching (n=1).

The data independently extracted from each study were the study population, demographics, clinical manifestations, diagnostic techniques, intra-left ventricular pressure gradients, associated disorders, nature of the accessory ventricular septum, treatment of choice, and patient outcomes.

SPSS software version 22.0 was used for statistical analysis. The measurement data were expressed in mean ± standard deviation and were compared by independent sample/paired t-test. The categorical variables were compared by Fisher exact test. $p<0.05$ was considered statistically significant.

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RESULTS

The 29 case reports, which dealt with a single patient in each, included in total 29 patients.[1,2,5-31] One case was a 24-week gestation fetus, whose gender was not reported.[29] 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),[2,6,11,13,16,17,19,28-30] and 19 (65.5%) adult patients (χ^2=5.59, p=0.035).[1,5,7-10,12,14,15,18,20-27,31]

Fourteen patients (48.3%) were asymptomatic,[2,6,8,13,14,16,17,22-24,26,27,29,31] while 15 (51.7%) were symptomatic (χ^2=0.00, p=1.000).[1,5,7,9-12,15,18-21,25,28,30]

Among 14 asymptomatic patients, 6 (42.9%) were pediatrics (including the fetal case), and 8 (57.1%) were adults (χ^2=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,[2,18,25,31] and in 9 (69.2%) patients, a systolic murmur was heard (χ^2=3.85, p=0.115).[1,5,6,11,12,21,28,30]

The locations of the murmurs were: left sternal border (n=4),[1,11,12,30] apex (n=2),[21,28] base (n=1),[5] mesocardiac area (n=1),[6] and unspecified (n=1).[23]

In 20 pateints, their electrocardiographic (ECG) findings were available: the ECG findings were normal in 6 (30%) patients.[2,6,25,26,29,31] In comparison, an abnormal ECG was seen in the remaining 14 (70%) patients (χ^2=6.40, p=0.026). In the 6 patients with a normal ECG, 3 (50%) were pediatric and 3 (50%) were adult cases (χ^2=0.00, p=1.000); while 3 (21.4%) patients with an abormal ECG finding were pediatrics and 11 (78.6%) were adults (χ^2=9.14, p=0.007). The abnormal electrocardiographic findings were heterogeneous, such as left bundle branch block,[15,18] atrial fibrillation,[20] and left anterior hemiblock,[9] *etc*.

All patients were initially inspected by transthoracic echocardiography, by which signs of DCLV could be visible as, a finger-like projection muscle band,[23] pronounced trabeculations interspersed by deep recesses,[22] flows in the intertrabecular recesses,[8] thinning of the left ventricular wall and the inter-ventricular septum,[10] and aneurysm-like structure.[15] But in 2 patients, the DCLV was primarily diagnosed as ventricular septal defect,[24] and ventricular and atrial septal defects,[2] respectively.

Nine (9/29, 31.0%) patients have one or more congnital heart defects, and 19 (19/29, 65.5%) pateints were associated with other cardiac abnormalities (Table I).

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

Congenital heart defect
1. VSD (perimembrane+muscular), Eisenmenger symdrome, TR, mitral valve prolapse, MR, DCRV (n=1)[5]
2. VSD (multiple, muscular, swiss-cheese), ASD, left circumflex artery with an anormal origin from the proximal RCA (n=1)[7]
3. VSD (multiple, muscular, swiss-cheese) (n=1)[17]
4. Subaortic obstruction (tunnel-type), mitral valve prolapse (n=1)[10]
5. Partial atrioventricular canal (n=1)[23]
6. Subaortic obstruction (n=1)[11]
7. Aortic stenosis (n=1)[19]
8. ASD (n=1)[30]
9. LV non-compaction (n=1)[8]
Secondary cardiac anomalies
1. LVOTO (n=11)
2. Myocardial scarring/fibrosis (n=4)
3. Coronary lesion (n=10)
4. Hypertension (n=8)
5. Pulmonary artery hypertension (n=3)
6. Diffuse subendocardial fibrosis (n=1)[10]
7. Cardiac necrosis markers (+) (n=1)[25]
8. Coronary embolism with subsequent ST-segment elevation myocardial infarction and transient ischemic attack (n=1)[9]
9. IVS hypertrophy (n=1)[26]
10. LV hypertrophy, RV contractile aneurysm, MR, TR (n=1)[12]
11. Apical RV aneurysm, right atrial dilation, RV hypertension (n=1)[11]

ASD: atrial septal defect; DCRV: double-chambered right ventricle; IVS: interventricular septum; LV: left ventricle; LVOTO: left ventricular outflow tract obstruction; MR: mitral regurgitation; RCA: right coronary artery; RV: right ventricle; TR: tricuspid regurgitation; VSD: ventricular septal defect.

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

Accessory septum	n (%)
A muscle band[2,6,7,8,10,15,16,20,22,23,26]	11 (55)
A membranous structure[1,13,19,27]	4 (20)
A fibromuscular ridge[11,21,28]	3 (15)
Prominent trabeculations[9,14]	2 (10)

Besides, 2 (2/29, 6.9%) patients had genetic problems: MYH7 rare variant,[28] and Vacterl association.[2]

The dimensions of the accessory chamber of the left ventricle was (described as long axis×short axis, mm^2) measured as 1898,[27] 1392,[25] 1020,[18] 2000,[14] and 378,[30] with a mean of 1337.6 ±666.6 mm^2. A comparison between those of the adult,[14,18,25,27] and that of the pediatric patients,[30] showed a significant difference (1577.5 ±228.5 mm^2 *vs*. 378 mm^2, p=0.011). An adult patient was reported to have an end diastolic volume of the accessory left ventricle of 38 mL/m^2.[22] The size of intra-left ventricular communication was 1.2 ±0.8 cm (n=7). Flow velocity across the accessory ventricular septum was 1.1 ±0.3 m/s during systole,[1,29,31] and was 1.0 ±0.3 m/s during diastole.[29,31] The intra-left ventricle pressure gradient before treatment was reported for 8 patients: it was zero in five patients,[2,6,12,13,28] (it rose to 55 mmHg after exercise in one of them[12]). Their mean pressure gradient was 30.1 ±46.2 mmHg (n=8).

The pressure gradient decreased to 14.3 ±16.9 mmHg (n=3) after treatment.[11,19,21] The post-treament pressure

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

Feature	Aneurysm	Diverticulum	DCLV
Communicating opening[4,15]	Wide neck	Narrow neck	By the anomalous septum or muscle bundle
Wall motion[4,15]	Dyskinetic	Absence of dyskinetic motion	Normal or dyskinetic
Wall structure[4,15]	Lack of full layers	Full layers	Full layers
Abnormal muscular and/or fiber bundles to division[29]	No	No	Yes
Associated congenital mid-line defects[22]	Seldom	Often	Seldom

DCLV: Double-chambered left ventricle.

gradient was much lower than that of before treatment, but lack of a statistical significance (30.1 ±46.2 mmHg *vs*. 14.3 ±16.9 mmHg, p=0.588).

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,[19] or an incomplete structure.[20,28] Their course was horizontal in several cases,[1,19] and was Y-shaped in a patient.[16]

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.[14] Besides, mangement of 14 (14/29, 48.3%) patients was mentioned: 11 (78.6%) were conservatively and 3 (21.4%) were surgically treated (χ^2=9.14, p=0.007).

The surgical indications of the 3 pateints undergoing a surgical treatment were an intra-left ventricular pressure gradient of 110 mmHg,[11] a high flow velocity of >2 m/s,[1] and severe left ventricular outflow obstruction with necessity of concurrent Ross procedure.[19] 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 (χ^2=0.29, p=1.000) and between pediatric and adult patients (χ^2=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.[17] 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.[15,32]

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.[29] 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.[32] 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.[32]

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.[15] When symptomatic or when associated with other cardiac abnormalities, surgical treatment is recommended.[33]

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.

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