# Clinical analysis on application of color Doppler echocardiographic in cortriatrium diagnosis

D.-L. CHEN<sup>1</sup>, X.-J. AN<sup>2</sup>, C.-Y. CUI<sup>3</sup>, M. QIN<sup>1</sup>, J.-X. DU<sup>1</sup>

**Abstract.** – OBJECTIVE: Discuss the main points of diagnosis of cortriatrium; patient's color Doppler echocardiography (CDE), provide basis for clinical treatment.

PATIENTS AND METHODS: Inspect 12 cortriatrium cases with CDE, 10 cases with cardiovascular angiography, 12 patients were confirmed by operation. Operations were all carried out under the moderate hypothermia cardiopulmonary bypass with intracardiac correction technique. Abnormal diaphragm in the left a trial was completely removed, and other combined heart malformations were also cured.

RESULTS: Four cases for II A type, 1 case for II B type, 6 cases for II A type, 1 case for II B type. Among them, there were 7 cases for combined atrial septal defect, 5 cases for ventricular septal defect, 3 cases for patent ductus arteriosus, 6 cases for pulmonary arterial hypertension. Twelve children all survived, deformity correction was satisfactory, and after operation, recovery went on well in 6 months to 3 years.

CONCLUSIONS: CDE has specific diagnostic value for cortriatrium; thus, it is the optimal method of diagnosing cortriatrium.

Key Words:

Cortriatriatum, CDE, Transportation mouth, Cardiopulmonary bypass, Cardiac arrest.

# Introduction

Cortriatrium is a rare congenital heart disease, with the incidence rate about 0.1%-0.4% of congenital heart diseases<sup>1-2</sup>. Typical cortriatrium is in the left atrial. We have cured 12 cases of cortriatrium with operations. The following is the report. Discussion concerning the diagnosis and surgical treatment of cortriatrium color Doppler echocardiographic (CDE) have been made with a review of the literature.

#### **Patients and Methods**

Twelve cases of cortriatrium confirmed by operation of our hospital were collected; among

them, there were 7 males, 5 females, aged from 6 months to 14 years old, and the average age is 6.2. Philips iE 33 type color Doppler ultrasonography (Amsterdam, The Netherlands) was adopted, with the probe frequency of 3.5-5.0 MHz. Supine or left lateral position was adopted by patients. Firstly, two-dimensional ultrasound was used to scan parasternal long axis view, apical four-chamber view, parasternal short-axis view, and subxiphoid four-chamber view. We mainly observed whether there was abnormal echo of diaphragm within the atrium, whether there was transportation mouth between real atrium and accessory atrium, and whether there was any other deformity. Next, type-M ultrasonography was adopted to measure the diameters of all the heart chambers. Finally, color Doppler flow imaging (CDFI) was used to observe whether there was color blood-flow signal at the transportation mouth. CDE features were compared with angiocardiography and operation, 10 cases with cardiovascular angiography, and all the cases were confirmed by operation. Median sternotomy incision was applied in operations under the conditions of low temperature and extracorporeal circulation cardiac arrest. Myocardial protection was accomplished by pouring cold crystalloid cardioplegia into aortic root and local ice cooling for heart. Left atrial inner structure was explored by atrial septal defect or cutting the atrial septal. Abnormal diaphragm in left atrial was completely removed. There were 10 cases of cortriatriumdiaphragm with hole, and 2 cases without hole. Treatment was made at the same time of combined malformations.

## Results

As can be seen in sonograph, according to Bank classification method, among the 12 cases of cortriatrium, there are 4 cases of IA type, and

<sup>&</sup>lt;sup>1</sup>Department of Ultrasonography, Xuzhou Children's Hospital, Xuzhou, Jiangsu, P.R. China

<sup>&</sup>lt;sup>2</sup>Department of Cardiology, Xuzhou Children's Hospital, Xuzhou, Jiangsu, P.R. China

<sup>&</sup>lt;sup>3</sup>Department of Cardiothoracic Surgery, Xuzhou Children's Hospital, Xuzhou, Jiangsu, P.R. China



**Figure 1.** Abnormal septum in the left atrium shown by apical four-chamber view.

the accessory atrium is connected with part of the pulmonary vein. Two-dimensional ultrasound displays abnormal echo of diaphragm of the left atrial in the apical four-chamber view (Figure 1), and the left atrium is divided into 2 chambers. Accessory atrium and real atrium are connected with a transportation mouth. It can be seen from CDFI that colorful shunts (mainly red) flow into the real atrium at the transportation mouth of apical four chamber view (Figure 2). One case belongs to IB type, with accessory atrium and part of the pulmonary vein connected, while accessory atrium and the left atrium disconnected. Blood of accessory atrium flows into the right atrium first, and then connects with the real atrium via atrium transportation. It can be seen from CDFI that there is no blood flowing between accessory atrium and real atrium, but there is blood flowing through atrial septum. Six cases belong to II A type, the accessory atrium is connected with 4 branches of pulmonary vein. Two-dimensional ultrasound displays abnormal echo of diaphragm of the left atrial. Accessory atrium and real atrium are connected with a transportation mouth. It can be seen from CDFI that colorful shunts (mainly red) in accessory atrium flow into the real atrium through the transportation mouth. One case belongs to II B type, with the accessory atrium connected with 4 branches of pulmonary vein, while the accessory atrium and the left atrium disconnected. Blood of accessory atrium flows into the right atrium first, and then connects with the real atrium via atrium transportation. It can be seen from CDFI that there is no blood flowing between accessory atrium and real atrium, but there is blood flowing through atrial septum. As can be seen in CDFI, for 4 cases of combined ventricular septal defect, there are colorful shunts (mainly red) from left to right at the ventricular septal in apical four-chamber view. And for 3 cases of combined patent ductus arteriosus, there are dual-phase shunts from left to right at the right pulmonary artery in aortic short axis view. For 4 cases of combined pulmonary artery hypertension, regurgitation pressure of three tricuspid valve port is measured with continuous wave (CW) Doppler to calculate the pulmonary artery systolic pressure.

After operation, all the patients achieved automatic heart re-beating, breaking away from cardiopulmonary bypass successfully. Eight patients were sent to Intensive Care Unit (ICU), ventilator and vasoactive drugs were used, and tracheal intubation was removed after stopping the adoption of medication and ventilator. Among them, 1 patient was complicated by right pneumothorax at the night of operation, and after the closed drainage of the right thoracic cavity, extubation was made the second day after operation. Four elder patients whose weights were more than 15 kg did not use ventilator and vasoactive drugs after operation, and successfully removed the tracheal intubation after a period of time when sent back to ICU. All the patients fully recovered and discharged from hospital. In the following 6 months~3 years after operation, the growth of all the patients has been normal or improved without other complications.

# Discussion

It refers to abnormal membranous structure in the left atrium, which divides the left atrium into 2 cavities. Thus cortriatrium has 3 cavities, the right atrium is normal, the left atrium is composed of real left atrium (the proximal left atrial) and accessory atrium (the distal left atrium)<sup>3,4</sup>. It is caused by abnormal growth of primary septum or the failure of mixture of pulmonary vein and the left atrium in the process of embryonic devel-



**Figure 2.** Color blood flow from accessory atrium to real atrium through transportation mouth shown by CDFI.

opment<sup>5,6</sup>. The accessory atrium is located in the upper right side of left atrium, and it is connected with pulmonary vein<sup>7</sup>. The cavity which is connected with the left ventricle through mitral valve is the real cavity. There may be or may not be transportation mouth between real atrium and accessory atrium<sup>8</sup>.

Bank is classified into type I (part type) and type II (complete type), according to pulmonary venous return degree received by accessory atrium; it is further classified into 2 subtypes according to whether there is transportation mouth between real atrium and accessory atrium: type A (connected with real atrium, with transportation), type B (disconnected with real atrium, without transportation). Currently, Bank classification is mostly adopted, because it is in accordance with the basic methods and rules of intracardiac inspection in surgery<sup>9,10</sup>. Key points of ultrasound cardiogram diagnosis:

- 1. Linear or membrane-shaped high echo in the left atrium fibromuscular membrane seen in two-dimensional ultrasound is the specific representation of cortriatrium diagnosis<sup>11</sup>. Sword four-chamber section comments on the connection between pulmonary vein and accessory atrium about whether there is transportation mouth between real atrium and accessory atrium and whether the interatrial septum is complete. According to literature<sup>12</sup>, if one end of the diaphragm echo is from the posterior wall of the left atrium, while the other end is attached to posterior aortic wall, then such case is complete cortriatrium. But if one end of the diaphragm is from the posterior wall of the left atrium, while the other end is not attached to posterior aortic wall (attached to the left atrial wall instead), then, such case is likely to be part cortriatrium<sup>13</sup>;
- 2. CDFI can observe whether there is color blood flow in the transportation mouth between real atrium and accessory atrium, in the atrial septal or in the ventricular septal. Cortriatrium should be identified with the narrow ring on mitral valve. The narrow ring is usually located horizontally with valve ring, while the septum of cortriatrium is distant from mitral valve ring; diaphragm ring on mitral valve is below the oval fossa and left atrial appendage, which is nearly attached to the root of mitral valve and can influence the opening of mitral valve. Cortriatrium septum is located above the two, which can be easily identified by multisection observation.

The key to the operation lies in careful inspection during the process. The inspection should abide by the order of extracardiac prat → right atrium → left atrium, clearly identifying the four pulmonary vein openings, the left atrial appendage, and mitral valve structure. And whether there is combined cardiovascular malformation should also be inspected. When cutting off the attachment part of lateral wall of the left atrial septum, be careful not to cut too deep in order to avoid breaking into the atrium wall or the coronary sinus; completely remove membranous tissues so as to avoid the diaphragm remnants prevent pulmonary venous blood flowing into the heart. After removing the septum, inspect again whether there is obstruction between real atrium and accessory atrium, whether four pulmonary vein openings are all connected with the posterior wall of the left atrium, and whether the mitral valve is complete, meanwhile, completely cure other combined heart malformations.

As can be seen from the above materials. among all the typical cortriatrium cases, complete type with transportation between real atrium and accessory atrium is the most common. Because the natural prognosis of cortriatrium is not ideal, no matter which type it is, if there is no transportation or the diameter of transportation mouth is less than 3 mm, the early mortality rate is high, and once cortriatrium is diagnosed, operation should be implemented as soon as possible<sup>14</sup>. The key to the operation lies in the careful inspection during the process, carefully identifying anatomic relationship, completely cutting off abnormal septum in the left atrium, and meanwhile cure intracardiac malformation. In recent years, CDFI has nearly taken the place of invasive inspection such as angiocardiography and cardiac catheterization in the diagnosis of cortriatrium<sup>15</sup>.

#### **Conclusions**

CDE plays an important role in the regular follow-up phase after cortriatrium operation, with its advantages of non-invasiveness, rapidity, simplicity, convenience, good repeatability, etc., it has become the preferred method of diagnosing cortriatrium and postoperative follow-up.

#### **Conflict of Interest**

The Authors declare that they have no conflict of interests.

## References

- YONGCHANG Z, WANXUE G. Ultrasonic medical science. The Fifth Edition. Beijing: Science and Technology Press; 2006: 252-255.
- CARBONE R, BOSSONE E, BOTTINO G, MONSELISE A, RUBENFIRE M. Secondary pulmonary hypertensiondiagnosis and management. Eur Rev Med Pharmacol Sci 2005; 9: 331-342.
- VIKHE VB, BORAWAKE K, GUPTA A, JAIN J. Cortriatrium presenting as mitral stenosis and severe anemia. N Am J Med Sci 2013; 5: 155-156.
- 4) ABRAHEEM A, POTTER L. Cortriatrium in Klippel-Feil syndrome. Eur J Intern Med 2009; 20: e27-28.
- ZENGWEI W, WEIYONG L, BAOREN Z. Collected works of operation-cardiovascular surgery. Beijing: People's Medical Publishing House, 1995; pp. 353-356.
- XIA J, ZHIYAN L. Clinical ultrasound diagnosis of pediatric heart disease. Wuhan: HubeiScience and Technology Press, 1998; pp. 263-268.
- TRAN HJ, GORDON R, ALLOGGIAMENTO T, NAGI SK, KRISH-NASWAMI A. Atrial fibrillation and cortriatriatum sinister: a case report. Perm J 2015; 19: e105-106.
- SABNIS GR, UDARE AS, PHADKE MS, KERKAR PG. Classic cortriatriatum with opposing shunts at the atrial level: a rare association. Eur Heart J 2015; 16: pii: ehv099.
- TAKEUCHI Y, KUROGANE K, NISHIMURA Y, KAJIURA T, NAKATA H. Asymptomatic cor triatrium in an elderly

- patient-observation by biplanar transesophageal Doppler echocardiography. Jpn Circ J 1997; 61: 189-191.
- FUKAZAWA M, ORITA H, HIROOKA S, GOTOH S, WASHIO M. A rare case of total anomalous pulmonary venous connection to the right atrium confounding with cortriatrium. Kyobu Geka 1994; 47: 295-298.
- FUJIMATSU T, TSUNEMOTO M, SHIMADA M, OHTA Y, ENDO S, ISHIZAWA A, KOIKE K, AKAGI M, WATARI T, KOBAYASHI J. Subtotal cortriatrium with left partial anomalous pulmonary venous connection-successful correction in an infant. Rinsho Kyobu Geka 1990; 10: 76-80.
- YAN P, LIU HY. Cortriatriatum: diagnostic echo cardiographic features. J Cardiovascular Tech 1991; 10: 1.
- LYNGBORG K, ANDERSEN M, EFSEN F. Transseptal left heart catheterization andleft atrial angiocardiography in the diagnosis of cortriatrium. Scand J Thorac Cardiovasc Surg 1970; 4: 149-152.
- 14) ALPHONSO N, NORGAARD MA, NEWCOMB A, UDEKEMY, CHRISTIAN P, COCHRANE A. Cortriatriatum: presentation, diagnosis and long-term surgical results. Ann Thorac Surg 2005; 80: 1666-1671.
- 15) Hui L, Min F, Fu-Min L, Lei S, Peng L, Jian Z. Transvaginal three-dimensional color power Doppler ultrasound and cervical MVD measurement in the detection of cervical intraepithelial neoplasia. Eur Rev Med Pharmacol Sci 2014; 18: 1979-1984.