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# Surgical Treatment of Neonatal Cantrell Pentalogy: A Case Report and Literature Review

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Neonate; Cantrell; Pentalogy; Surgical treatment

## 1. Abstract

**1.1. Objective:** to explore the pathophysiology, prenatal and postnatal diagnosis, treatment, surgical methods and prognosis of Cantrell pentalogy.

**1.2. Methods:** The clinical data of a neonate with Cantrell pentalogy were analyzed retrospectively, and the related literatures were reviewed.

**1.3. Results:** The clinical and imaging features of the children were consistent with the diagnosis of Cantrell's pentalogy. Three surgical operations were performed in the neonatal period, including end-to-side anastomosis of aortic arch under cardiopulmonary bypass, patch repair of ventricular septal defect, repair of atrial septal defect, ligation of arterial catheter, correction of cardiac vascular malformation and repair of chest and abdominal wall defect, and the postoperative recovery was in good condition.

**1.4. Conclusion:** Cantrell pentalogy is a rare congenital dysplasia, and its condition is complex. Surgery is an effective treatment method, and the prognosis is related to complicated malformation.

## 2. Introduction

Pentalogy of Cantrell (PC) is a collection of hereditary malformations invoking the heart, pericardium, diaphragm, sternum, and ventral abdominal wall [1]. This disease is complicated in condition, low in survival rate and difficult to treat. Recently, a PC patient was admitted to Children's Hospital affiliated to Soochow University. Three surgical operations were performed during the neonatal period, and all deformities were corrected with satisfactory results. The report is as follows.

## 3. General Information

The child was G1P2, the youngest twin, born at 37+1W, born with a weight of 2400g and an Apgar score of 10. Prenatal cardiac ultrasound indicated that he suffered from ventricular septal defect, atrial septal defect and aortic arch dysplasia. Due to the influence of twins, the structure of anterior abdominal wall is unclear. Physical examination: The lips are slightly cyanotic, the lower end of sternum is suspiciously missing, the apex beating point is located at the lower edge of the median xiphoid process of sternum, and the upper abdominal muscle below the beating part is defective, the skin is still intact and dark red, and the cord shape seems to be connected with the umbilicus who (Figure1A). Heart sounds are still strong, and IV/VI grade heart murmurs can be heard. Auxiliary examination: cardiac ultrasound and cardiac CTA prompt: Cardiac dextration and ortic coarctation with aortic arch dysplasia, ventricular septal defect, atrial septal defect, patent ductus arteriosus(PDA) and persistent left superior vena cava. Chest CT and 3D reconstruction showed that the sternum was ossified incompletely and the lower end of sternum was missing. (Figure. 2)(Figure. 3). Prostaglandin was given to keep the arterial catheter open after admission, and endotracheal intubation and ventilator assisted ventilation were given 9 days after birth.

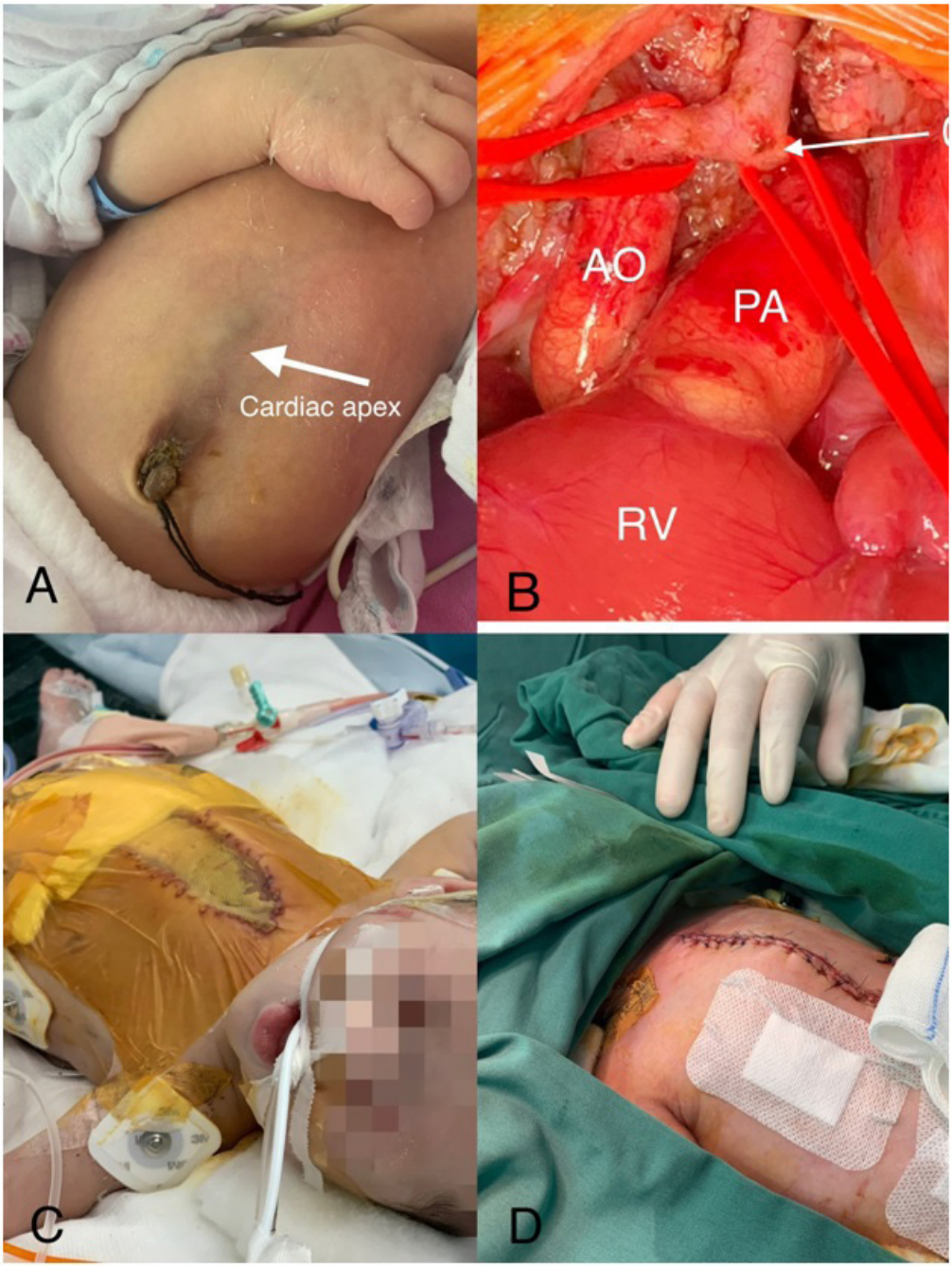


**Figure 1:** Note:
A chest and abdomen appearance and apex position.
B aortic arch anatomy.
C Delayed chest closure after the first operation.
D Chest and abdomen incision after three operations.

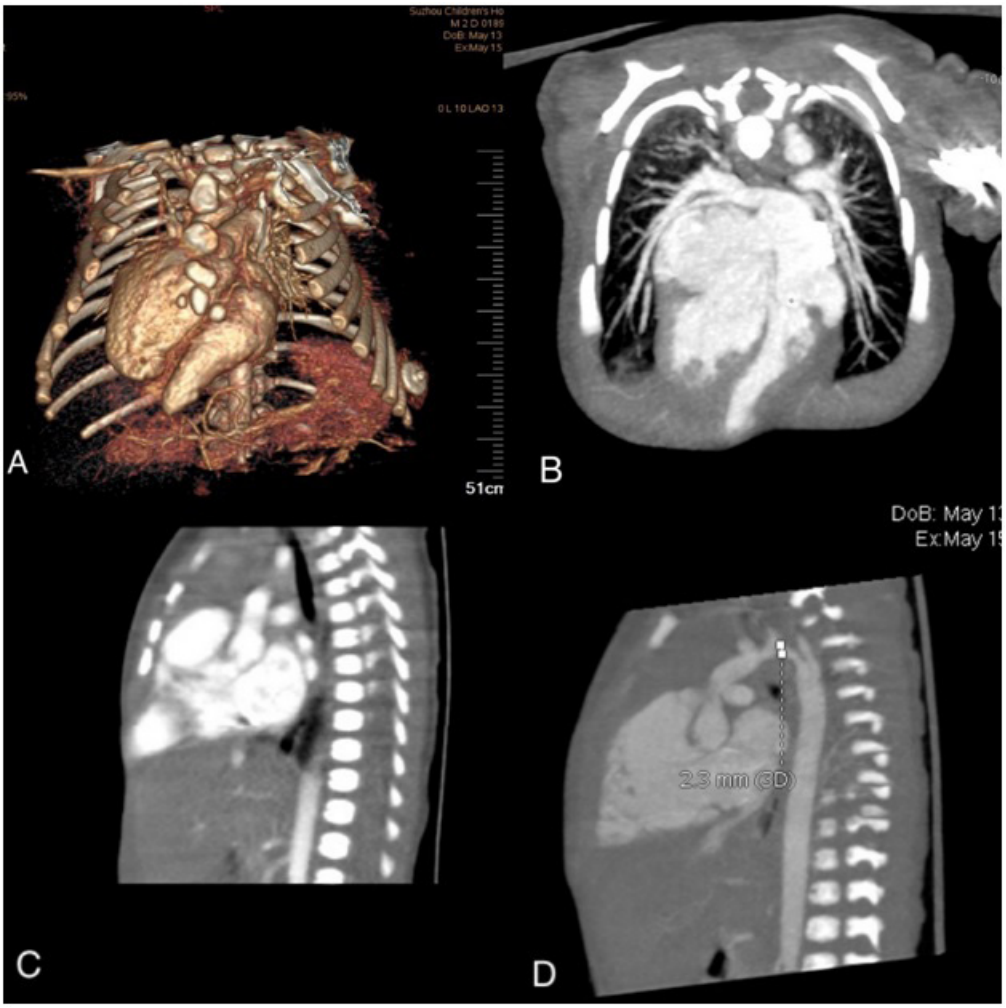


**Figure 2:** Note:
A enhanced cardiac CT and three-dimensional reconstruction of sternum and ribs indicate incomplete sternum ossification, and the lower sternum is missing.
B enhanced cardiac CT cross section indicates that the lower sternum and muscular layer of abdominal wall are defective. The heart protrudes forward.
C enhanced cardiac CT oblique sagittal position shows that the apex moves down to the abdomen.

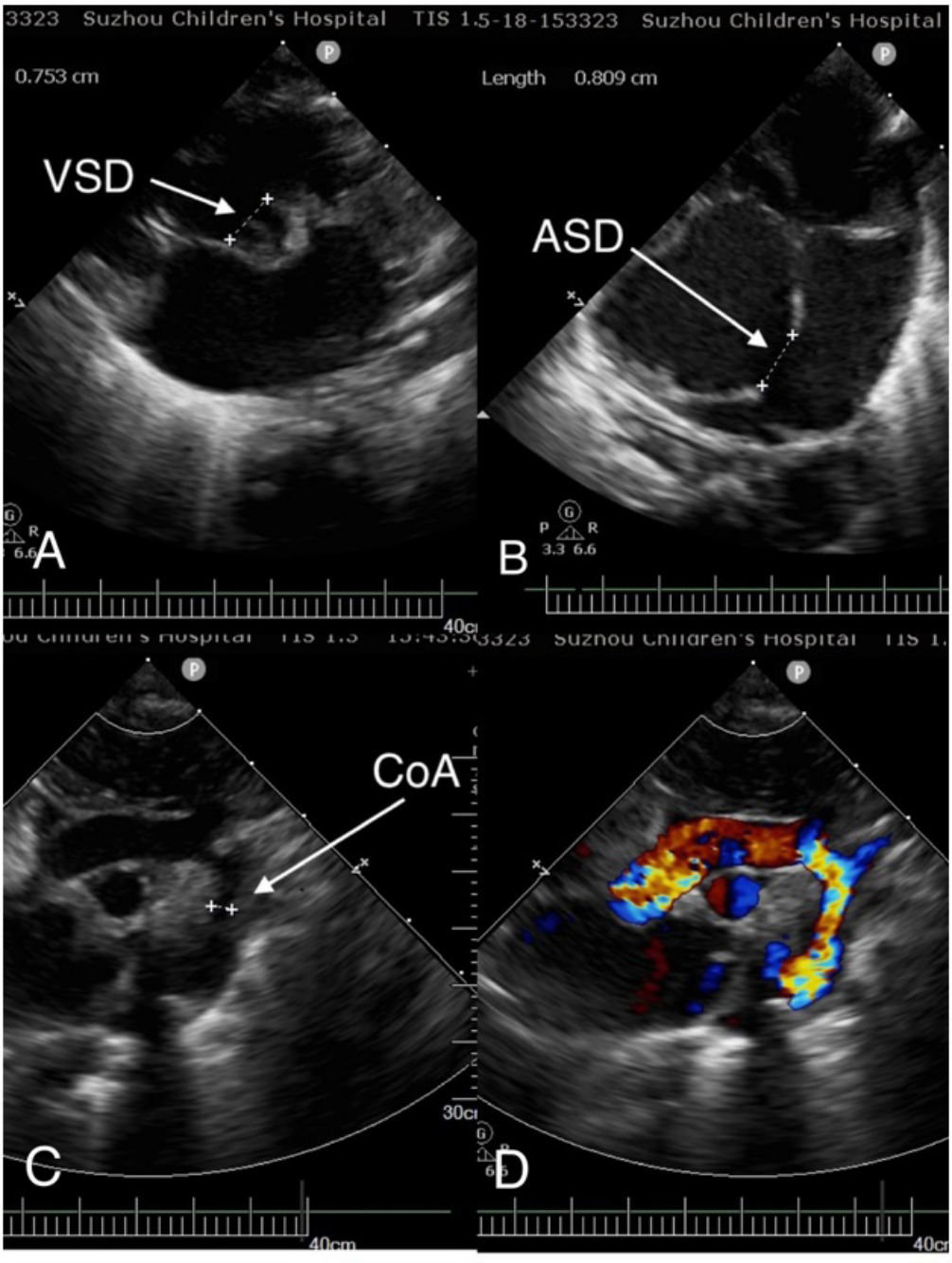


**Figure 3:** Note:

A short axis section of super-large heart artery, ventricular septal defect.

B four-chamber heart section shows atrial septal defect.

C transverse dysplasia of aortic arch with constriction.

D color Doppler indicates that the blood flow velocity in aortic arch, especially in aortic isthmus, is accelerated.

**4. Operation Condition**

On the 12th day after birth, the first operation was performed, including end-to-side anastomosis of aortic arch, patch repair of ventricular septal defect, repair of atrial septal defect and ligation of arterial catheter under deep hypothermic circulatory arrest (DHCA+CACP). During the operation, it was found that the lower sternum was defective, the rectus abdominis muscle was defective, and the diaphragm was intact, the attachment point of its front edge moved down to the navel, and the anterior lower wall of parietal pericardium was defective, forming a capsular bag with apex adhered. Dysplasia of aortic arch, which is about 3-4mm straight and light near the distal end, and the isthmus of aorta is obviously narrowed, with an inner diameter of only 1-2mm. The artery is close to closure. There are bilateral superior vena cava with small diameters. The diameter of large perimembranous ventricular septal defect is 8mm, Atrial septal defect 8mm.The median sternal incision was taken and extended down to the navel to dissociate the adhesion between apex and capsular bag. The ascending aorta

and right atrium cannula were used to establish cardiopulmonary bypass, and the arterial catheter was ligated and cut off. When the rectal temperature was 25 degrees, blocked the ascending aorta, and perfused HTK myocardial protection solution to result in cardiac arrest. Stop cardiopulmonary bypass, push the aortic cannula to the innominate artery and block it, and start anterograde cerebral perfusion. The flow rate is 30ml/kg. Thoroughly resect the isthmus of the aortic arch and the arterial catheter tissue, cut it along the long axis at the proximal bottom of the aortic arch, and perform end-to-side anastomosis with the cut descending aorta (7/0 prolene), stop the cerebral perfusion, retract and advance the aortic cannula to the anastomosed aortic arch, block the aorta again, restart the bypass, stop the circulation when the temperature drops to 18C, and pass through the right atrium.The ventricular septal defect was repaired with bovine pericardial biological patch 6/0 prolene, and the atrial septal defect was sutured with 6/0 prolene. Suture the right atrial incision, re-circulate with cardiopulmonary bypass and re-warm. The cardioversion is sinus rhythm. Evacuated cardiopulmonary bypass, delayed chest closure and skin incision was sutured with Silastic patch. Total time of cardiopulmonary bypass: 177min; Aortic occlusion time: 102min; Circulation stopping time: 43+30min, anterograde cerebral perfusion time: 34min. Adrenalin and ramodulin was given after operation. Seventeen (17) days after birth, the edema of the child has relieved, and the second operation was performed, including diaphragm folding and abdominal muscle repair. Twenty-one (21) days after birth, after the third operation, sternal suture was completed and chest closure was performed. (Figure 1BCD). Child recovered gradually after the operation and has been discharged from hospital. After 1 and 3 months of follow-up and was in good condition.

## 5. Discussion

PC is a rare congenital malformation, which was first reported by Cantrell in 1958 and revised by Toyama in 1972 [2]. PC can be diagnosed by ultrasound in early pregnancy [3]. Prenatal diagnosis helps families to make an informed decision on whether to continue pregnancy. It is extremely rare for twins to have PC [4]. In this case, one of the twins has PC, and the other fetus is healthy.‑Cantrell's definition of PC mainly involves the following five aspects, including intracardiac structural malformation, right-lateral heart defect, diaphragmatic pericardium defect, lower sternum defect, defect in front of diaphragm and midline of upper abdomen [1 ] [5]. Because it is often complicated with cardiac heterotopia, it is also called thoracic and abdominal cardiac heterotopia. Patients can show the existence of all five defects, or they may only have some defects. It can be divided into complete type and incomplete type [6] [7]. Defects also vary greatly in degree. For example, the defect of sternum may be manifested as complete loss of sternum, or it may be cracked or shorter than normal. The severity of the disease varies accordingly. In some cases, the heart protrudes beyond the chest and abdomen, as if the heart is beating outside the body, especially in patients with omphalocele and abdominal fissure [3]. There are many kinds of deformities in this child, which are consistent with the manifestation of complete PC.‑Compound malformation is the most common cardiac malformation, especially ventricular septal defect. Atrial septal defect, left ventricular diverticulum, pulmonary artery stenosis, tetralogy of Fallot, and double outlet of right ventricle are also frequently reported [8]. This child's cardiovascular malformation is characterized by severe aortic coarctation with arch dysplasia, which is rarely reported in literatures [8].‑The general consensus of PC treatment is to make individualized plan according to the complexity of chest and abdominal wall defect and intracardiac malformation. The primary management of newborn babies is particularly important. The strategies include neonatal resuscitation, keeping hemodynamic stability, and possibly needing ventilation support, inhaling nitric oxide(NO) and extracorporeal membrane oxygenation. Prostaglandins may also be needed to maintain the patency of arterial catheters in order to treat catheter-dependent congenital heart disease. Surgical treatment is an important method for radical treatment of PC. Many methods of reconstruction and repair have been described in the literature. The surgical purposes include correcting cardiac malformation, restoring cardiac position and anatomical structure, and repairing defects of chest and abdomen wall and diaphragm.‑The mortality rate of patients with cardiac heterotopia was as high as 52% [9],The main causes were cardiac rupture, tamponade, cardiac arrest, endocarditis, vascular embolism, heart failure and arrhythmia [3][9]. The prognosis of patients with severe complete PC is poor, the average survival rate without surgical intervention is about 36 hours [3].‑We believe that staged operation is more appropriate, which is consistent with the literature report [7]: If the congenital heart disease of PC patients is not so serious as to pose no threat to life, then the early surgical intervention is mainly to repair the abdominal wall muscles and diaphragm, so as to let the heart return to the chest first, and if the heart structure is abnormal, we can judge whether or when to perform the operation according to the results of long-term follow-up. However, this child was diagnosed with aortic arch constriction with dysplasia and large ventricular septal defect. Due to the gradual closure of the arterial catheter, the blood flow in the descending part of the aortic arch is reduced, and the left ventricular pressure is continuously increased, which leads to the rapid occurrence of heart failure in children. Therefore, it is necessary to complete the correction of cardiac malformation as soon as possible. Similar to conventional heart surgery, Correction of cardiac malformation is carried out under cardiopulmonary bypass. Because of ectopic heart and lack of pericardium in front of the heart, the heart and its blood vessels often adhere to surrounding tissues, so care should be taken not to damage coronary vessels when separating.Because the child is light in weight, only 2500g during the operation, and there are bilateral superior vena cava, which makes it difficult to

intubate the vena cava, the technique of deep hypothermic circulatory arrest was adopted during the operation, and the anterograde cerebral perfusion technique was adopted during the aortic arch reduction, thus ensuring the aortic arch forming effect and completely repairing the ventricular septal defect. Because of postoperative myocardial edema and other reasons, the strategy of delaying chest closure was adopted, and the abdominal muscles were repaired by the second operation and the diaphragm was folded, which also created conditions for the heart to completely return to the chest cavity in the next step. Finally, all deformities were corrected and good results were achieved.The complications of PC range from surgery to anesthesia, and the lack of timely intervention will further complicate the patient's condition. In addition, complications of cardiac malformation repair surgery and postoperative intra-abdominal and intra-thoracic hypertension may lead to cardiac decompensation and intra-abdominal organ damage. Therefore, an interdisciplinary team is needed to manage such patients. The team should consists of obstetricians, neonatologists, radiologists, pediatric cardiologists, pediatric cardiac surgeons or general surgeons [10]. Thereby reducing the occurrence of complications to the greatest extent and improving the survival rate of patients.

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