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Operative Technique

# Surgical treatment and outcomes of pentalogy of Cantrell in eight patients

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ABSTRACT

*Purpose:* To summarize the experience and feasibility of radical one-stage surgical repair of pentalogy of Cantrell in eight cases in a single tertiary care center.

*Patients and Methods:* From August, 2007 to June, 2011, eight patients with pentalogy of Cantrell were diagnosed. One of them had undergone intracardiac operation 2 years ago, leaving a large thoracoabdominal defect with most of the heart outside the chest. After correction of cardiac malformations, patients were individually treated with different operative procedures to restore their cardiac position and anatomy and repair their thoracoabdominal and diaphragmatic defects.

*Results:* Surgical procedures in all the patients were uneventful. Seven cases underwent one-stage complete correction. The only patient candidate for second-stage operation underwent extended thoracoplasty because of inadequate accommodation for the heart. Diaphragmatic defects were repaired with a PROCEED patch in two cases and closed directly in the remainder. The right sixth and seventh ribs were grafted for chest reconstruction in two cases and the right seventh rib in one case. Chest wall clefts were closed directly in the remaining five cases. A left ventricular diverticulum was removed in one of the two patients. An interventricular residual shunt (0.5 cm) was observed in one case and treated by transcatheter closure on the fifth day after operation. One case with double outlet right ventricle suffered from low cardiac output syndrome during the postoperative period and recovered after medical treatment. The early and midterm clinical outcomes were satisfactory without death or serious complications during the follow-up period of 24–72 months.

*Conclusion:* The diversity and complexity of POC result in a variable clinical course, with different clinical manifestations which require unique surgical procedures. Innovative multidisciplinary management strategies are necessary to achieve better outcomes. The key points in surgical treatment are: a) to obtain satisfactory correction of the cardiac malformation, b) adequate space for return of the heart to the mediastinum, and c) a thoracoabdominal wall repair which protects the heart from excessive compression or distortion. If treated correctly, most patients with less complex cardiac defects who survive the newborn period can be successfully treated via a one-stage operation.

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Pentalogy of Cantrell (POC), firstly described in 1958 by Cantrell et al. [1], is a rare syndrome which is characterized by the presence of five major malformations: midline supraumbilical abdominal wall defect, lower sternal defect, diaphragmatic pericardial defect, anterior diaphragmatic defect and various congenital abnormalities of the heart [2,3], with an incidence of 5.5–7.9 per million [4]. Because of the severity and complexity of the disease, few patients survive through early childhood. Surgical treatment is complex and a wide variety of operative techniques are utilized, often requiring a staged approach [5–7]. Here we report successful one-staged radical correction in seven cases and a staged repair in one case.

## 1. Patients and methods

### 1.1. Patients

This retrospective review was completed with institutional review board approval. Between August, 2007 and June, 2011, 8 patients were admitted to Children's Hospital of Qingdao University (ages ranged from 4 months to 26 years: mean age 7.48 years). There were 6 males and 2 females. The first patient was admitted for second-staged (plastic) repair who had undergone repair of an intracardiac malformation 2 years ago at another institution, leaving a large thoracoabdominal defect with most of the heart in an extrathoracic location. The successful operation was reported on our national TV and wide disseminated on Internet. In a short period seven cases who had a complete manifestation of POC syndrome (a midline

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supraumbilical abdominal wall defect, a defect of the lower sternum, a deficiency of the anterior diaphragm, a defect of the diaphragmatic pericardium and different congenital intracardiac defects) came from all over our country to our hospital seek surgical treatment. All patients, to varying degrees, exhibited incomplete ectopia cordis (EC) with the heart (covered by pigmented, loose skin) beating outside the thoracoabdominal or thoracic wall, especially in the standing position. The heart could generally be partially or completely repositioned into the thoracic cavity when the patient was in a supine position. Seven were classified as thoracoabdominal type and one as a thoracic type. Complex intracardiac anomalies were found in one case and dynamic pulmonary hypertension (PH) in two cases. A persistent left superior vena cava (LSVC) was found in six cases, draining to the right atrium through its usual location of the coronary sinus in all (additional patient characteristics are shown in Table 1).

### 1.2. Methods

The incision was made through median sternotomy from angulus sterni to the lower end of abdominal defect. Care was taken neither to injure the unprotected heart (especially coronary artery or it's important branches) nor to break the peritoneum when separating adhesions between the subcutaneous tissues and cardiac surface. The whole heart and thoracoabdominal defect were exposed completely. It was possible to see the separated rectus sheath under the adipose layer attached to the lower ribs. Any intracardiac anomaly was firstly corrected on cardiopulmonary bypass. Subsequently, about 1.5–2 cm anterior to the phrenic nerve (taking care to avoid injury to the phrenic nerve), bilateral pericardium and pleurae were completely opened to expand the space for heart restoration. Then a trial of restoring the heart to the chest was performed under close monitoring. Tentatively and gradually, push the heart back into the thoracic cavity, observing changes in heart rate, blood pressure and rhythm for at least 10 minutes. If it could be completely repositioned with no hemodynamic changes, both osteal edges of the chest wall defect were gradually drawn together. More extensive plastic chest reconstructive procedures were individualized when necessary. Patients with a relatively small chest wall defect whose heart can be repositioned easily with minimal or no hemodynamic changes, were selected to have defect directly closed after bilaterally amputating the 6–7 costal cartilages. The bony edges of the defect were approximated with stainless steel wire (Fig. 1). Patients with a large segmental chest wall defect whose heart could be repositioned easily had the defect repaired using autologous material.

For patients without adequate accommodation space for heart, a more complex extended thoracoplasty technique was adopted. Grafts for chest wall reconstruction were selected from the right 6–7 ribs, which were harvested through the same incision, positioned transversely, and securely sutured with stainless steel wires to both sides of the thoracic defect forming a cage over the heart (Fig. 2). Similarly, the diaphragmatic defects were closed directly or repaired with PROCEED patch (Johnson & Johnson). This is a partially biodegradable membrane produced which provides strong abdominal wall support and antiadhesion effects as well as good biocompatibility. It is often used for hernia and abdominal wall defect repairs. A PROCEED patch was also chose as pericardial substitute to further protect the heart as well as reduce tissue adhesion in patients who require potential repeat sternotomy. The abdominal wall defect was repaired by interrupted sutures approximating the rectus sheaths.

## 2. Results

Case 1 (Fig. 3), a child who had undergone intracardiac malformation repair 2 years ago was admitted for second-staged (plastic) repair. Computerized tomographic angiography (CTA) showed a large lower sternal defect with most of the heart outside the chest, with about 2/3 of the heart in front of the liver under the diaphragm. During operation, a large segment defect about 8 × 12 cm in anterior diaphragm was found behind the heart. Part of the colon and liver herniated through the diaphragmatic defect into the chest.

**Table 1**
Summary of patients and postoperative outcomes.

| Patient (No.) | Gender (M/F) | Age (years) | Cardiac malformation | Other accompanied complication | Chest wall defect Td × D (cm) | Diaphragmatic defect Td × D (cm) | Postoperative complication |
|---|---|---|---|---|---|---|---|
| 1[a] | F | 7 | Double outlet right ventricle<br>Ventricular septal defect<br>Left superior vena cava | Diaphragmatic hernia[b] | 8 × 10 | 8 × 12 | No |
| 2 | M | 10 | Ventricular septal defect<br>Pulmonary hypertension (moderate)<br>left superior vena cava | Cleft lip | 6 × 8 | 3 × 5 | Residual shunt (0.5 cm) |
| 3 | F | 26 | Atrial septal defect | No | 5 × 6 | 3 × 5 | No |
| 4 | M | 4 | Ventricular septal defect<br>Left ventricular diverticulum | No | 8 × 6 | 3 × 4 | No |
| 5 | M | 4 | Double outlet right ventricle<br>Ventricular septal defect<br>Atrial septal defect<br>Pulmonary stenosis<br>Left superior vena cava | No | 4 × 5 | 2 × 4 | Low cardiac output syndrome |
| 6 | M | 8 | Ventricular septal defect<br>Atrial septal defect pulmonary<br>Hypertension (severe)<br>Left superior vena cava | Bilateral bullae of lung | 5 × 7 | 3 × 5 | No |
| 7 | M | 6/12 | Ventricular septal defect<br>Atrial septal defect<br>Left superior vena cava | No | 3 × 4 | 1.5 × 4 | No |
| 8 | M | 4/12 | Ventricular septal defect<br>Atrial septal defect<br>left ventricular diverticulum<br>Left superior vena cava | Cleft lip | 4 × 5 | 2 × 3 | No |

Td: transverse diameter of the defect; D: diameter of the defect.
[a] Intracardiac malformations have corrected previously.
[b] Refers to that part of the colon and liver herniated through the diaphragmatic defect into thoracic cavity.

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