Simultaneous repair of pectus excavatum and congenital heart disease over the past 30 years

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Received 21 March 2002; received in revised form 18 August 2002; accepted 2 September 2002

Abstract

Objective: Pectus excavatum may be present in patients requiring operations for cardiac defects. The study was undertaken in order to assess our simultaneous repairs of pectus excavatum and congenital heart disease over the past 30 years. Methods: Between 1970 and 2000, 12 patients underwent simultaneous repair of pectus excavatum and congenital heart disease. Six of 12 patients had ventricular septal defects as cardiac malformations (subgroup A). Operative technique, after the intracardiac procedure using cardiopulmonary bypass, consists of total subperichondrial resection of deformed costal cartilages, transection of deformed portion of the sternum in 2–3 points, and fixation of the sternum in elevated position using two Kirschner wires and a bridge external traction. Postoperative catheterization was performed in five of 12 patients (subgroup B). We evaluated the operative data, the improvement of pectus deformity and right ventricular performance retrospectively. The operative data in subgroup A were compared with those in recent random patients with ventricular septal defects only or with pectus excavatum only (control groups).

Results: There was no operative death and non-serious complications were seen in nine patients (atelectasis in six, superficial wound infection in two, chylothorax in one). Pectus deformities improved with the drop of vertebral index postoperatively. The mean total operative time and postoperative drainage in subgroup A were 128.4% and 123.7%, respectively of those in the ventricular septal defect control group. The mean perioperative bleeding in subgroup A was more than the sum of those in control groups. Right ventricular end-diastolic (RVEDVI), end-systolic (RVESVI), stroke (RVSVI) volume indices and ejection fraction (RVEF) in subgroup B tended to increase after surgery. In particular, there were significant increases of RVEDVI (35%, \( P < 0.05 \)) and RVSVI (77%, \( P < 0.01 \)). Conclusion: Simultaneous cardiac and pectus repairs were performed successfully without serious complications. Moreover, simultaneous repair resulted in an improvement of right ventricular performance with significant increases of RVEDVI and RVSVI.

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Keywords: Simultaneous repair; Pectus excavatum; Congenital heart disease; Vertebral index; Right ventricular performance

1. Introduction

Pectus excavatum is the most common congenital chest wall malformation and is usually seen as an isolated congenital abnormality. However, it is occasionally associated with cardiac abnormalities, such as Marfan’s syndrome or congenital heart disease [1–4]. Surgical correction of pectus excavatum is mainly performed for cosmetic and physiological reasons. In addition, cardiopulmonary complications from mechanical compression by the depressed chest wall may be an account to perform surgical correction. Especially, the patients with underlying cardiac disease may have cardiopulmonary complications by the presence of the chest wall depression. Cardiac compression can contribute to postoperative hemodynamic instability if the pectus deformity is left uncorrected [5]. We reviewed our experience with simultaneous repairs of pectus excavatum and congenital heart disease over the past 30 years to investigate the propriety of our surgical policy to these deformities.

2. Material and methods

2.1. Patients

Between 1970 and 2000, 12 patients underwent simultaneous repair of pectus excavatum and congenital heart disease. There were five male and seven female patients, and their ages at operation ranged from 10 months to 11 years. Diagnoses of congenital heart disease included ventricular septal defect (\( n = 6 \)), atrial septal defect (\( n = 2 \)), tetralogy of Fallot (\( n = 2 \)), double outlet right...
ventricle \( (n = 1) \) and complete atrioventricular canal defect \( (n = 1) \). Postoperative catheterization was performed in five of 12 patients. We defined two subgroups for retrospective analyses. Six patients with ventricular septal defect were defined as subgroup A. Five patients with postoperative catheterization were defined as subgroup B (Table 1).

### 2.2. Operative technique

Operation is started by median sternotomy, and the repair of cardiac abnormality is undertaken in the usual fashion using cardiopulmonary bypass. After the intracardiac procedure and sternal closure using wires of surgical steel, the repair of pectus deformity is carried out. The pectoral muscle flaps are elevated off the sternum and costal cartilage. Total subperichondrial resection of the deformed costal cartilages is achieved by incising the perichondrium anteriorly, and usually the entire third to eighth cartilages are removed to the costochondral junctions bilaterally. The deformed portion of the sternum is transected in two or three points, and the divided sternum is fixed and corrected in elevated position by using two Kirschner wires. The opened perichondrium is resutured. In eight cases since 1980, we applied additional bridge external traction for about 10 days after operation [6] (Fig. 1).

### 2.3. Evaluation of pectus deformity

To determine the severity of the deformity in all patients, we measured three indices of sternal depression on the lateral chest radiographs (Fig. 2): (1) the vertebral index \( B/A \) of Fallot; DORV, double outlet right ventricle; CAVC, complete atrioventricular canal defect. Subgroup A has VSD as associated congenital heart disease \( (n = 6) \).

- **Table 1**

<table>
<thead>
<tr>
<th>Congenital heart disease</th>
<th>No. of patients ( b )</th>
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<tbody>
<tr>
<td>VSD</td>
<td>6 (1)</td>
</tr>
<tr>
<td>ASD</td>
<td>2 (0)</td>
</tr>
<tr>
<td>TF</td>
<td>2 (2)</td>
</tr>
<tr>
<td>DORV</td>
<td>1 (1)</td>
</tr>
<tr>
<td>CAVC</td>
<td>1 (1)</td>
</tr>
<tr>
<td>Total</td>
<td>12 (5)</td>
</tr>
</tbody>
</table>

\( a \) VSD, ventricular septal defect; ASD, atrial septal defect; TF, tetralogy of Fallot; DORV, double outlet right ventricle; CAVC, complete atrioventricular canal defect. Subgroup A has VSD as associated congenital heart disease \( (n = 6) \).

\( b \) Number of patients with postoperative catheterization in parentheses.

![Fig. 1. Operative technique. (a) Operation is started by median sternotomy. After the intracardiac procedure and sternal closure using wires of surgical steel, the perichondrium of the bilateral 3rd to 8th costal cartilages are opened longitudinally and the deformed cartilages are removed subperichondrially. The depressed portion of the sternum is transected in 2-3 points. (b) The divided sternum is fixed and corrected in elevated position using 2 penetrating Kirschner wires inserted in a parallel fashion. The lower end of the Kirschner wire is suspended by a heavy nylon suture applied as a sling and pulled out of the skin and hooked on the external bridge.](https://academic.oup.com/ejcts/article-abstract/22/6/874/445309)

![Fig. 2. Indices assessing the amount of the sternal depression (lateral view of the chest X-ray).](https://academic.oup.com/ejcts/article-abstract/22/6/874/445309)
sternovertebral distance), (2) D/C ratio (maximum sagittal depth of depression of sternum/distance from anterior surface of the vertebral body to the sternum at Louis’s angle) and (3) E/C (minimal sagittal distance from anterior surface of the vertebral body to the sternum). We compared these indices before and after operation. The improvement of pectus deformity is indicated by a drop in vertebral index and D/C ratio, and an elevation in E/C ratio after operation.

2.4. Operative data

The operative outcome was evaluated by total operation time, perioperative bleeding, postoperative drainage, duration of ventilation and complications. The operative data in subgroup A were compared with 50 recent random patients with ventricular septal defect (VSD) only (VSD control group) and 50 patients recent random patients with pectus excavatum only (pectus control group) who have undergone surgical repairs in our institution.

2.5. Evaluation of right ventricular performance

In subgroup B, the right ventricular performance was evaluated with pre-/postoperative comparison of end-systolic (RVESVI), end-diastolic (RVEDVI) and stroke volume indices (RVSVI), and ejection fraction (RVEF).

2.6. Statistical analysis

All data are expressed as mean ± SD. Preoperative and postoperative right ventricular performance were compared by a two-tailed t-test to identify statistically significant differences. A P-value of less than 0.05 was considered statistically significant.

3. Results

3.1. Respect to operative variables

Characteristics and operative data in a total of 12 patients are presented in Table 2. Postoperative complications were as follows: atelectasis in six patients, superficial wound infection in two, and chylothorax in one. Three patients with atelectasis required reintubation. Patients with wound infection required debridement, irrigation, drainage and antibiotic therapy because of a suspicion of mediastinitis. However, cultures of discharge or soft tissue from the mediastinum were all negative. The follow-up period ranged from 3 to 25 years with good outcomes. There were no operative deaths and no late complications. No development of pectus deformity required reoperation.

3.2. Evaluation of pectus deformity

Measurements of sternal depression on lateral chest radiographs are detailed in Table 3. Preoperative vertebral index (B/A), D/C and E/C ratio were 0.28 ± 0.12, 0.39 ± 0.15, and 1.20 ± 0.54, respectively. Humphreys [8] classified pectus excavatum on the vertebral index and the age. On this basis, preoperative vertebral indices were ‘mild’ in eight patients, ‘moderate’ in three patients, and ‘severe’ in one patient. Up to 1 year after operation, results were ‘normal’ in nine patients, ‘mild’ in three patients. Postoperative vertebral index, D/C and E/C ratio were 0.25 ± 0.06, 0.26 ± 0.09 and 1.43 ± 0.38, respectively. There were drops in vertebral index and D/C ratio, and an elevation in E/C ratio after operation. These changes indicated the improvement of pectus deformities.

3.3. Evaluation of operative data in subgroup A

The mean total operative time and postoperative drainage in subgroup A were 128.4% and 123.7%, respectively, of those in the VSD control group, which were less than the sum of those in VSD and pectus control groups. But the mean perioperative bleeding in subgroup A was more than the sum of those in VSD and pectus control groups. Durations of ventilation in subgroup A was slightly longer than those in the VSD control group (Table 4).

3.4. Evaluation of right ventricular performance in subgroup B

The average values of the pre-postoperative RVEDVI, RVESVI, RVSVI and RVEF in subgroup B are shown in Fig. 3. RVEDVI increased significantly after surgery on average about 35% (P < 0.05). The average of RVESVI and RVEF after surgery increased, but the changes were not statistically significant. RVSVI increased significantly after surgery on average about 77% (P < 0.01).

### Table 2

<table>
<thead>
<tr>
<th>Age (mean ± SD)</th>
<th>10 months–11 years (5.6 ± 3.0)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex (n)</td>
<td>Male (5), female (7)</td>
</tr>
<tr>
<td>Weight (kg)</td>
<td>6.5–30.0 (16.9 ± 7.5)</td>
</tr>
<tr>
<td>Total operation time (min)</td>
<td>335–610 (470 ± 89)</td>
</tr>
<tr>
<td>CPB time (min)</td>
<td>57–268 (144 ± 65)</td>
</tr>
<tr>
<td>ACC time (min)</td>
<td>0–173 (61 ± 45)</td>
</tr>
<tr>
<td>Perioperative bleeding (ml/kg)</td>
<td>6.2–63.5 (29.0 ± 19.5)</td>
</tr>
<tr>
<td>Postoperative drainage (ml/kg)</td>
<td>12.7–56.4 (30.6 ± 14.0)</td>
</tr>
<tr>
<td>Complications (n)</td>
<td>Atelectasis (6), Superficial wound infection (2), Chylothorax (1)</td>
</tr>
</tbody>
</table>

* CPB, cardiopulmonary bypass; ACC, aortic cross-clamp.
4. Discussion

Charles and colleagues [9] reported a 26-year experience with pectus deformity repairs, including nine cases (11.6%) with simultaneous intracardiac repair. In our review, the incidence of congenital heart defects in patients with pectus deformity repair was 5.5%. A review from the Children’s Hospital in Boston documented a 0.17% incidence of pectus deformities in children with congenital heart defects [10]. The incidence of pectus deformity repair in patients with congenital heart defects was 0.5% in our institution.

A wide variety of surgical techniques for correction of pectus deformity have been described [11–15]. Our technique is based on sternocostal elevation. From 1970 to 1979, a total of 29 children with pectus deformity, including four children with simultaneous intracardiac repair, were operated by total subperichondrial resection of deformed cartilages (usually third to eighth, bilaterally), transection of deformed portion of the sternum in two or three points, fixation of the sternum in elevated position using two Kirschner wires, and resuture of the opened perichondrium. The use of Kirschner wires provides rigid fixation of the sternum, resulting in earlier union with primary osseous healing [16]. However, a child with severe adenoid had paradoxical breathing and dyspnea due to flail chest after the operation. The trial of utilizing bridge external traction brought a dramatic clinical improvement in this patient. Since this experience in 1980, we applied an addition of the bridge external traction for about 10 days after the operation of pectus deformity [6]. The combination of the bridge external traction improved early postoperative outcome in sternocostal elevation with Kirschner wires. Chest wall instability with paradoxical movement could lead to pulmonary complications and require prolonged intubation in patients already having some compromise of pulmonary function after a major cardiac procedure [17,18].

The problems presented by the pectus excavatum deformity are difficulty in accurately dividing the sternum in the midline and retraction symmetrically. But our routine operative exposure obtained from median sternotomy approach was excellent. It was also enough for one patient with the most severe deformity in our series to access the heart and to accomplish the open cardiac procedure.

We have evaluated the severity of pectus deformity on three indices of sternal depression on lateral chest X-ray film. The vertebral index is most applicable in evaluating pectus deformity, which is considered the deformity in relation to the patient’s age [8,19]. Recently we have also adopted a pectus index to assess the severity of pectus deformity, which can be derived from dividing the transverse diameter of the chest by the anterior–posterior diameter on a simple computed tomography scan [20].

When the heart defect is repaired but the pectus deformity is not addressed, the compression of the heart by pectus deformity may severely compromise the hemodynamics after the intracardiac repair [5]. Some authors suggest a two-stage procedure [9,21]. Jones and colleagues [1] recommend a two-stage approach, correcting the pectus deformity several months before the cardiac pathology. They point out that one-stage procedures may prolong the duration of the operation, may increase the risk of bleeding, and may not improve the exposure to the heart. But the correction of pectus deformity before the cardiac pathology, we are afraid, may have a bad influence upon wound healing in severely cyanotic children. We agree with a two-stage procedure if the heart defect is associated with high morbidity, but we believe if this is not the case that one-stage procedure can be performed without added incidence of

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Table 4
Operative data in subgroup A and control groups

<table>
<thead>
<tr>
<th></th>
<th>Subgroup A (n = 6)</th>
<th>Control group</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>VSD (n = 50)</td>
<td>Pectus (n = 50)</td>
</tr>
<tr>
<td>Total operation time (min)</td>
<td>443 ± 95</td>
<td>345 ± 69</td>
</tr>
<tr>
<td>Perioperative bleeding (ml/kg)</td>
<td>24.1 ± 14.6</td>
<td>14.3 ± 11.4</td>
</tr>
<tr>
<td>Postoperative drainage (ml/kg)</td>
<td>31.8 ± 26.6</td>
<td>25.7 ± 11.2</td>
</tr>
<tr>
<td>Duration of ventilation (days)</td>
<td>3.2 ± 3.1</td>
<td>2.6 ± 3.9</td>
</tr>
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*a* VSD, ventricular septal defect.

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Fig. 3. Right ventricular performance before and after surgery. The average of RVEDVI and RVSVI increased significantly after surgery. RVEDVI, right ventricular end-diastolic volume indices; RVESVI, right ventricular end-systolic volume indices; RVSVI, right ventricular stroke volume indices, RVEF, right ventricular ejection fraction.
complications and problems. Charles and colleagues [9] reported that patients with congenital heart defects can undergo simultaneous cardiac and pectus deformity repair, with no added morbidity and excellent long-term results with regard to chest wall contours. In our clinical review, the increase of operative time and postoperative drainage in subgroup B were only 28.4 and 23.7% of those in the VSD control group. On the other hand, perioperative bleeding in simultaneous repair tends to increase because of the necessity of full heparinization on cardiopulmonary bypass. But simultaneous repairs were performed on the most recent two patients with no blood transfusion.

Surgical repair of pectus deformity brings cosmetic and functional improvement. Fonkalsrud and colleagues [15] have proved that more than 90% of patients have improvement in exercise tolerance, endurance, and cardiac and respiratory symptoms after operation. Before the operation, the patient with pectus deformity usually is significantly different from normal subjects in terms of right ventricular function because right ventricular anatomy is altered by sternal compression of the right atrium and right ventricle [22,23]. Kowalewski and colleagues described that surgical repair of funnel chest causes a significant increase of RVEDVI, RVESVI, and RVSFI [24]. In our reviews, RVEDVI and RVESVI importantly increased after simultaneous repair of pectus excavatum and congenital heart disease. We are aware of the limitations in evaluating the right ventricular function clearly, because postoperative right ventricular volume is reflected not only by the release from the cardiac compression by the sternum but also by the release from a volume or a pressure overload on the right ventricle.

We conclude that our simultaneous repairs of pectus excavatum and congenital heart disease were performed successfully, with good improvement of pectus deformity. There was no serious complication. In addition, simultaneous repair resulted in a significant increase of RVEDVI and RVESVI.

References