Cardiac Surgery in Cleft Lip and Cleft Palate Children: Srinagarind Hospital Experience

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Background: Congenital heart disease is one of the common incidents associated with craniofacial anomalies.

Objective: To date, there are no published studies from Thailand on review experience regarding operation of congenital heart disease in cleft lip/cleft palate patients. This study aims to report the patterns of our practices in those patients.

Material and Method: A hospital-based, descriptive study during 1996-2013 in congenital heart disease associated with cleft lip/cleft palate in Srinagarind Hospital, Faculty of Medicine, Khon Kaen University, Khon Kaen Province, Thailand.

Results: During the period of study there were 2,600 cases of cleft lip/cleft palate. Congenital heart disease was found in 60 patients (2.3%). Only 11 patients had heart surgery performed, 7 patients were operated on by open heart surgery, 2 PDA (Patent Ductus Arteriosus) ligation and 2 Blalock-Tausig shunts. The most common perioperative complication was aspirated pneumonia. No 30-day mortality occurred.

Conclusion: Congenital heart disease in cleft lip and cleft palate patients is not uncommon. A multidisciplinary team approach is the key to success in management. Heart operation can be performed at anytime if clinically indicated.

Keywords: Congenital heart disease, Cleft lip and cleft palate, Hospital mortality

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Table 1. Diagnosis of congenital heart disease confirmed by echocardiography before operation

<table>
<thead>
<tr>
<th>Age</th>
<th>Weight</th>
<th>Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 year</td>
<td>9 kilogram</td>
<td>TOF</td>
</tr>
<tr>
<td>4 months</td>
<td>3,250 gram</td>
<td>PDA, ASD, VSD</td>
</tr>
<tr>
<td>1 months</td>
<td>3,250 gram</td>
<td>SV, dTGA, AVSD, PA, PDA</td>
</tr>
<tr>
<td>24 days</td>
<td>2,640 gram</td>
<td>PDA, VSD, ASD</td>
</tr>
<tr>
<td>1 year</td>
<td>6 kilogram</td>
<td>DORV, VSD, PDA</td>
</tr>
<tr>
<td>2 months</td>
<td>2,800 gram</td>
<td>PS, PDA</td>
</tr>
<tr>
<td>11 years</td>
<td>35 kilogram</td>
<td>ASD secundum</td>
</tr>
<tr>
<td>1 year</td>
<td>7 kilogram</td>
<td>VSD, PDA</td>
</tr>
<tr>
<td>12 years</td>
<td>3,500 gram</td>
<td>Sub aortic AS, bicuspid AV, aberrant Rt. subclavian artery</td>
</tr>
<tr>
<td>2 months</td>
<td>3,830 gram</td>
<td>TGA, VSD, PDA, ASD</td>
</tr>
<tr>
<td>6 months</td>
<td>3,800 gram</td>
<td>ASD primum</td>
</tr>
</tbody>
</table>

TOF = tetralogy of fallot; PDA = patent ductus arteriosus; ASD = atrial septal defect; VSD = ventricular septal defect; SV = single ventricle; dTRA = dextro-transposition of great artery; AVSD = atrioventricular septal defect; PA = pulmonary atresia; DORV = double-outlet of right ventricle; PS = pulmonary stenosis; AS = aortic stenosis; AV = aortic valve

Table 2. Closed heart surgery patients

<table>
<thead>
<tr>
<th>Patients</th>
<th>Diagnosis</th>
<th>Operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>TOF</td>
<td>Lt. MBT stunt</td>
</tr>
<tr>
<td>2.</td>
<td>PDA, ASD, VSD</td>
<td>PDA ligation</td>
</tr>
<tr>
<td>3.</td>
<td>SV, dTGA, AVCD, PA, PDA</td>
<td>Lt. MBT shunt</td>
</tr>
<tr>
<td>4.</td>
<td>PDA, VSD, ASD</td>
<td>PDA ligation</td>
</tr>
</tbody>
</table>

TOF = tetralogy of fallot; PDA = patent ductus arteriosus; ASD = atrial septal defect; VSD = ventricular septal defect; SV = single ventricle; dTRA = dextro-transposition of great artery; AVSD = atrioventricular septal defect; PA = pulmonary atresia; DORV = double-outlet of right ventricle; PS = pulmonary stenosis; AS = aortic stenosis; AV = aortic valve

Table 3. Data of open heart surgery children

<table>
<thead>
<tr>
<th>Patient number</th>
<th>Diagnosis</th>
<th>Operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>5.</td>
<td>DORV, VSD, PDA</td>
<td>VSD patch + PDA ligation</td>
</tr>
<tr>
<td>6.</td>
<td>PS, PDA</td>
<td>Pulmonary valvotomy + PDA ligation</td>
</tr>
<tr>
<td>7.</td>
<td>ASD 2°</td>
<td>ASD patch</td>
</tr>
<tr>
<td>8.</td>
<td>VSD, PDA</td>
<td>VSD patch, PDA ligation</td>
</tr>
<tr>
<td>9.</td>
<td>Sub aortic AS</td>
<td>Subaortic resection</td>
</tr>
<tr>
<td>10.</td>
<td>TGA, VSD, ASD</td>
<td>Arterial switch operation</td>
</tr>
<tr>
<td>11.</td>
<td>ASD 1°</td>
<td>ASD patch + MV/TV repair</td>
</tr>
</tbody>
</table>

TOF = tetralogy of fallot; PDA = patent ductus arteriosus; ASD = atrial septal defect; VSD = ventricular septal defect; SV = single ventricle; dTRA = dextro-transposition of great artery; AVSD = atrioventricular septal defect; PA = pulmonary atresia; DORV = double-outlet of right ventricle; PS = pulmonary stenosis; AS = aortic stenosis; AV = aortic valve

130 minute (average 22.7 minute).
No 30 day hospital mortality occurred. All of the children had respiratory tract infection and usually aspirated pneumonia. Deep wound infection and recurrent laryngeal nerve injury was found in child number 5 (after PDA ligation) and patient number 6.
developed sub glottic stenosis after tracheostomy. The 3rd patient had BT shunt thrombosis, needing heparin infusion.

Many associated anomalies other than cleft lip and palate were found in some children such as club feet, esotropia. So they will need further operation or intervention to correct other defects later.

A multidisciplinary team composed of plastic surgeon, dentist, pediatrician, speech pathologist, ophthalmologist and special nurse are needed to collaborate caring for all patients.

Patient number 10 who was associated with Goldenhar syndrome, had many congenital anomalies. After he underwent arterial switch operation, closure VSD, PDA ligation and ASD repair when 2 months old, he then had performed check flab advancement for facial correction followed by cheiloplasty and palatoplasty at 1 year old. When two years old, ophthalmologists operated mid orbital advancement both sides. When he was four years old, he had performed total orbital advancement. The other physicians which attended him were ENT physician for ear function assessment, pediatrician for growth and development care and speech and language pathologist for speech management. All patients had experienced pneumonia often at birth or perioperative period.

Discussion

Cleft lip and palate are common craniofacial abnormalities seen in children. Congenital heart disease is usually associated with cleft lip and palate [6-10]. Cleft lip and cleft palate may be a genetically disorder [11-14]. We found 2.3% of congenital heart disease in cleft lip and cleft palate child in Khon Kaen University compared to 29% at Aga Khan University of Pakistan [15]. Birth order is a contributing factor in the origin of some isolated congenital anomalies [16].

We report 11 cases of congenital heart disease which were operated on in our unit. Four cases had performed closed heart surgery, 2 modified BT shunt and 2 PDA ligation. Seven cases were operated for open heart surgery. No 30 day mortality occurred. A common perioperative complication was aspirated pneumonia [17]. Because of difficulty of feeding and many associated anomalies, patients should be taken care of by a multidisciplinary specialist team [18].

They also receive long term follow-up for continuous care.

Conclusion

Heart surgery in cleft lip and palate children need care by a multidisciplinary team. A common perioperative complication was special aspirated pneumonia. Long term follow-up was important for their quality of life.

What is already known on this topic?

Congenital heart disease is a common anomaly associated with cleft lip and cleft palate patients.

What this study adds?

Experience of heart surgery in these patients.

Acknowledgement

This study was supported by Center of Cleft lip-Cleft Palate and Craniofacial Deformities, Khon Kaen University in association with the Tawanchai Project for publication.

Potential conflicts of interest

None.

References

ประสบการณ์ผ่าตัดหัวใจพิการแย่งเนื้อในเด็กปกคลุมเพศไหนของโรงพยาบาลศรีนครินทร์

สมคส พาบุญ, เดิดชัย คัลลีศรีศรี, หนึ่ง ปราโมช

กุศลพัฒนา: โรคหัวใจพิการแย่งเนื้อในเป็นความพิการที่พบรวมกันในประเทศไทย

วัตถุประสงค์: เพื่อศึกษาประมาณการและผลประโยชน์จากค่าของผ่าตัดหัวใจในเด็กที่มีความพิการปากแฉกพืายนิ้วรวมกับ

วัตถุประสงค์: การศึกษาแบบระดับมาตรฐานของผู้ดูแลโรคหัวใจพิการแย่งเนื้อใน คณะแพทยศาสตร์ มหาวิทยาลัยขอนแก่น ระหว่างปี พ.ศ. 2539-2556 ในการศึกษาโรคหัวใจพิการแย่งเนื้อในรวมกับความพิการปากแฉกพืายนิ้ว พบว่าช่วยคัดกรองโรคหัวใจ

ผลการศึกษา: เทียบกับความเป็นสาเหตุของโรคหัวใจ 2,600 ราย พบเป็นโรคหัวใจแย่งเนื้อใน 60 ราย คิดเป็นร้อยละ 2.3 มีค่าสูงกว่าเกณฑ์การคัดกรอง

11 ราย 7 ราย ผ่าตัดหัวใจแย่งเนื้อใน 4 ราย ผ่าตัดหัวใจแย่งเนื้อใน ภาวะแทรกซ้อนทั่วไปที่สูงคือ การคัดกรองโรคหัวใจจากการสังเกต ไม่มีการเสียชีวิต

หลังผ่าตัด 30 วัน

สรุป: การศึกษาโรคหัวใจพิการแย่งเนื้อในรวมกับความพิการปากแฉกพืายนิ้ว พบว่าช่วยคัดกรองโรคหัวใจพิการแย่งเนื้อ

สามารถทำได้ ในการศึกษาโรคหัวใจ в ประเทศศรีนครินทร์ ได้พบปัญหาทางคลินิกเกี่ยวกับคัดกรองโรคหัวใจจากการสังเกตได้อย่าง