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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Children with cleft lip and cleft palate commonly have associated congenital heart disease[1-4]. In Pakistan the risk of congenital heart disease in cleft children is about 23 times that of the general population compared to 16 times in Swedish population[5]. Congenital heart defect in cleft children is difficult to manage because of many associated anomalies and feeding difficulties.

Many aspects of treatment experience of congenital heart disease in cleft children should be reviewed to share experience.

Material and Method

A hospital-based descriptive study in cleft children during 1996-2013 who presented with congenital heart disease. All patients who were operated on for congenital heart disease were reviewed. Perioperative data such as diagnosis operation and immediate clinical outcome will be shown.

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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.

Results

Cleft lip and cleft palate patients numbered 2,600 cases during 1996-2013 in Srinagarind Hospital, Faculty of Medicine, Khon Kaen University, Thailand. Congenital heart disease was found in 60 patients (2.3%). Eleven patients of congenital heart disease underwent operation. All of this group were cleft lip and palate. Their diagnosis is shown in Table 1.

First group of patients, numbers 1,2,3,4 had performed closed heart surgery by 2 modified Blalock-Tausig shunt and 2 PDA ligation as show in Table 2.

The other 7 patients of numbers 5-11 were open heart surgical group, diagnosis shown and operation as Table 3.

Cardiopulmonary bypass technique was used with moderate systemic hypothermia, aortic cross clamp and blood carioplegia. CPB times were 23-236 minute. (average 63.7 minute) and aortic clamp times were 18-

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

	Age	Weight	Diagnosis
1.	1 year	9 kilogram	TOF
2.	4 months	3,250 gram	PDA, ASD, VSD
3.	1 months	3,250 gram	SV, dTGA, AVSD, PA, PDA
4.	24 days	2,640 gram	PDA, VSD, ASD
5.	1 year	6 kilogram	DORV, VSD, PDA
6.	2 months	2,800 gram	PS, PDA
7.	11 years	35 kilogram	ASD secundum
8.	1 year	7 kilogram	VSD, PDA
9.	12 years	3,500 gram	Sub aortic AS, bicuspid AV, aberrant Rt. subclavian artery
10.	2 months	3,830 gram	TGA, VSD, PDA, ASD
11.	6 months	3,800 gram	ASD primum

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

Patients	Diagnosis	Operation
1.	TOF	Lt. MBT stunt
2.	PDA, ASD, VSD	PDA ligation
3.	SV, dTGA, AVCD, PA, PDA	Lt. MBT shunt
4.	PDA, VSD, ASD	PDA ligation

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

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

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

Table 4. Associated disease of the patients (more than cleft lip, cleft palate and congenital heart disease)

Patients number	Associated disease, anomalies
2.	Di Gorge syndrome, Micrognathia, hypocalcaemia
3.	Bilateral undescended testis
5.	Bilateral club feet
6.	Right club feet
10.	Golden har syndrome

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[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 anomalities[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.

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Potential conflicts of interest

None.

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ประสบการณ์ผ่าตัดหัวใจพิการแต่กำเนิดในเด็กปากแหว่งเพดานโหว่ของโรงพยาบาลศรีนครินทร์

สมภพ พระธานี, เชิดชัย ตันติดิรินทร์, มนัส ปะนะมณฑา

ภูมิหลัง: *โรคหัวใจพิการแต่กำเนิดเป็นความพิการที่พบร่วมกับปากแหว่งเพดานโหว่*

วัตถุประสงค์: *เพื่อศึกษาประสบการณ์และผลลัพธ์เบื้องต้นของการผ่าตัดหัวใจในเด็กที่มีความพิการปากแหว่งเพดานโหว่ร่วมด้วย*

วัสดุและวิธีการ: *การศึกษาแบบบรรยายจากฐานข้อมูลของโรงพยาบาลศรีนครินทร์ คณะแพทยศาสตร์ มหาวิทยาลัยขอนแก่นระหว่างปี พ.ศ. 2539-2556 ในผู้ป่วยโรคหัวใจพิการแต่กำเนิดที่พบร่วมกับความพิการปากแหว่ง เพดานโหว่ ซึ่งมาผ่าตัดรักษาโรคหัวใจ*

ผลการศึกษา: *เด็กที่มีความพิการปากแหว่งและเพดานโหว่ 2,600 ราย พบเป็นโรคหัวใจแต่กำเนิด 60 ราย คิดเป็นร้อยละ 2.3 มีผู้ป่วยได้รับการผ่าตัดหัวใจ 11 ราย 7 ราย ผ่าตัดหัวใจชนิดเปิด 4 ราย ผ่าตัดหัวใจชนิดปิด ภาวะแทรกซ้อนที่พบบ่อยที่สุดคือ การติดเชื้อของปอดจากการสำลัก ไม่มีการเสียชีวิตหลังผ่าตัด 30 วัน*

สรุป: *ภาวะหัวใจพิการแต่กำเนิดที่พบร่วมกับความพิการปากแหว่ง เพดานโหว่ จะพบร่วมกันบ่อยกว่าคนปกติ การผ่าตัดรักษาหัวใจพิการแต่กำเนิดสามารถทำได้ ไม่มีการเสียชีวิตในโรงพยาบาล แต่พบปัญหาแทรกซ้อนหลังผ่าตัดเป็นปอดติดเชื้อจากการสำลักได้บ่อย*