# Coarctation of the Aorta in Children at Siriraj Hospital

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**Abstract**

A retrospective analysis of 33 children who were diagnosed to have coarctation of aorta at Siriraj Hospital between January 1989 and December 1998 was undertaken. There were 21 males (64%) and 12 females (36%). Their ages ranged from one day to 11 years (median 2 months). The majority of the patients (78.8%) were presented early within the first year of life. The predominant clinical manifestations were congestive heart failure (69.6%), systemic hypertension of the upper extremities (36.3%) and decreased femoral pulses. Chest roentgenogram revealed cardiomegaly (70%) and increased pulmonary blood flow (84%), reflecting congestive heart failure and associated left-to-right shunting. Electrocardiogram showed normal pattern (33.3%), right ventricular hypertrophy (33.3%), left ventricular hypertrophy (22.2%) and biventricular hypertrophy (11.2%). The younger the patient is, the more right ventricular predominance is demonstrated. According to the echocardiogram and/or aortogram, juxtaductal type was found in 51.5 per cent, postductal type in 27.3 per cent and preductal type in 21.2 per cent. Medical management included prostaglandin $E_1$ infusion in a newborn baby presented with low-cardiac output state, anticongestion and antihypertension, if indicated, followed by surgical correction. The result of coarctectomy with end-to-end anastomosis with or without arch augmentation was good. The operative mortality rate was 5 per cent. The overall mortality in the present study was 9 per cent. The most common causes of death were multiorgan failure and pulmonary infection. Residual coarctation was found in 5 per cent.

**Key word :** Coarctation of Aorta, Children

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Coarctation of the aorta (CoA), derived from Latin verb *arctare*, means to make tight. It is used to describe a narrowing aorta (Ao), most commonly in the thoracic descending Ao. True CoA results from a localized thickening of the aortic media, which protrudes from the posterior and lateral wall into the lumen of the vessel and obstructs blood flow[1].

It has been cited that 9 per cent of all children with congenital heart disease have some degree of CoA. As a dominant lesion, it ranks as the fourth most common cardiac defect causing symptoms in infancy (between 0.20 and 0.62 patients per 1000 livebirths) and accounts for 7.5 per cent of critically ill infants with heart disease[2]. At Siriraj Hospital, in 1997, we found CoA 2.1 per cent of all congenital heart defects, the twelfth rank[3]. As no clinical data of CoA in Thai children has ever been recorded, we would like to analyse our patients, compare to the Western study and imply our results to Thai patients.

## PATIENTS AND METHOD

All patients were diagnosed as having CoA with or without simple congenital heart defects, e.g., ventricular septal defects, bicuspid aortic valve, etc., by echocardiogram and/or cardiac catheterization at the Department of Pediatrics, Faculty of Medicine Siriraj Hospital between January 1989 and December 1998 and were retrospectively analysed. The exclusion criteria of the present study was CoA associated with complex congenital heart disease, e.g., single ventricle physiology lesion, transposition of the great arteries, etc.

Demographic data, clinical manifestations, chest roentgenogram, electrocardiogram including treatment and outcomes were evaluated.

## RESULTS

A total of 33 CoA patients, during the study period of 10 years, were analysed.

### Demographic data

The patients' age ranged from 1 day to 11 years (median 2 months). The weight ranged from 1.7 to 45.7 kg (mean 9.4 kg). The birth weight ranged from 1,750 to 4,980 g (mean 2,870 g). Twenty-one cases (64%) were male and 12 cases (36%) were female. The male: female ratio was 1.8:1.

### Clinical manifestations

The distribution of age at presentation is shown in Table 1. The clinical manifestations at initial were described in Table 2.

**Table 1. Age at presentation of patients (n = 33).**

| Age | Number (cases) | Frequency (%) |
|---|---|---|
| 0 - 1 month | 10 | 30.3 |
| 1 month – 1 year | 16 | 48.5 |
| 1 – 5 years | 1 | 3.0 |
| > 5 years | 6 | 18.2 |

**Table 2. The clinical manifestations at initial in the present study (n = 33).**

| Variable | Number (cases) | Frequency (%) |
|---|---|---|
| Symptoms | | |
| Tachypnea | 20 | 60.6 |
| Abnormal cardiac auscultation | 8 | 24.3 |
| Poor weight gain, poor feeding | 3 | 9.1 |
| Claudication | 1 | 3.0 |
| None* | 1 | 3.0 |
| Signs** | | |
| Congestive heart failure | 23 | 69.6 |
| Hypertension of upper extremities | 12 | 36.3 |
| Asymptomatic murmur | 8 | 24.2 |
| Low cardiac output | 2 | 6.6 |

* Incidentally, this patient was found to have systolic hypertension during acute hemorrhagic fever illness.
** One may have more than one clinical sign.

Only 28 cases (85%) were palpated four-extremity pulses at the first examination and all were able to be diagnosed CoA. In contrast to the remainders (5 cases), they were not palpated the four-extremity pulses and initially missed the diagnosis. Systolic murmur was heard in 31 cases (93.9%), continuous murmur in 1 case and no record in 1 case.

Systolic blood pressures between the upper and lower extremities were compared and demonstrated in Table 3. The majority (91%) had upper extremity pressure greater than or equal to the lower extremity one. In the hypertensive patients (12 cases, 36.3% of total cases), all were under 1 year old, except one. The systolic blood pressure ranged from 94 to 163 mmHg (mean 120 mmHg) and the diastolic pressure ranged from 57 to 97 mmHg (mean 60 mmHg). The distribution of age in these patients is shown in Table 4. All presented congestive heart failure, except one case who was incidentally discovered with hypertension during acute hemorrhagic fever illness.

**Investigations**

Chest roentgenogram revealed cardiomegaly (70%) and increased pulmonary vasculature (84%). Rib notching was found in 1 case, an 11-year-old boy. Electrocardiogram is described in Table 5. Echocardiogram and/or cardiac catheterization findings were classified into preductal, juxtaductal and postductal CoA. According to the classification, we compared our results to the Smallhorn's study[4] as shown in Table 6. Seven cases (21.3%) had mild CoA. Thirty-six per cent (7 out of 19 cases) had a transverse aortic arch diameter less than 50 per cent of the ascending Ao diameter. Isolated CoA was found in only 4 cases (12%). The majority were associated with congenital left-to-right shunt lesions.

**Table 3. The comparison of systolic blood pressure between upper and lower extremities (ext.) in the present study (n = 33).**

| Systolic blood pressure values | Number (cases) | % |
|---|---|---|
| Upper ext. = lower ext. | 6 | 18 |
| Upper ext. > lower ext. 10-20 mmHg | 5 | 15 |
| Upper ext. > lower ext. > 20 mmHg | 19 | 58 |
| Upper ext. < lower ext. (normal) | 3 | 9 |

**Table 4. Distribution of age in the systolic hypertension patients in the present study (n = 12).**

| Age | Range (Mean, mmHg) | Number (cases) | % |
|---|---|---|---|
| 0 – 1 month | 112-127 (119.5) | 2 | 16.6 |
| 1 month – 1 year | 94-138 (116.0) | 9 | 75.0 |
| > 1 year | 163 | 1 | 8.4 |

**Table 5. Electrocardiogram (ECG) findings in the present study (n = 33).**

| ECG pattern | Age | | Number (cases) | % |
|---|---|---|---|---|
| | Range | Mean | | |
| RVH | 1 day – 3 months | 1 month | 11 | 33.3 |
| LVH | 14 days – 11 years | 4 years | 7 | 22.2 |
| BVH | 1 day – 6 months | 2.8 months | 4 | 11.2 |
| Normal | 1 day – 11 years | 32 months | 11 | 33.3 |

RVH, right ventricular hypertrophy; LVH, left ventricular hypertrophy; BVH, biventricular hypertrophy

The associated congenital heart defects in the present study were compared to the Cobanoglu's study[5] (Table 7).

**Treatment (Table 8)**

Prosglandin $E_1$ was established in 2 cases, presenting low-cardiac output state and later, referred to surgery. Twenty cases (60%) underwent surgical repair. Eighty per cent of our cases performed coarctectomy with end-to-end anastomosis. Their mean age at surgery was 18.5 months (range 8 days to 14 years). There were 7 cases (35%) that underwent surgery at less than 3 months. Percutaneous transluminal catheter intervention, balloon angioplasty, was performed in 1 case of native CoA, with a good result.

**Outcomes and follow-up**

Twenty-three cases were regularly followed up, with the follow-up period ranging from 3 months to 10 years (mean 2.8 years). Seven cases were lost

**Table 6. The comparison of the coarctation type between the present study and the Smallhorn and associates' study[4].**

| Type | The present study (n = 33) | | Smallhorn, et al (n = 48) | |
|---|---|---|---|---|
| | no. | % | no. | % |
| Preductal | 7 | 21.2 | 40 | 83.3 |
| Juxtaductal | 17 | 51.5 | 5 | 10.4 |
| Postductal | 9 | 27.3 | 3 | 6.3 |

**Table 7. The associated congenital heart defects in the present study (n = 29), compared to the Cobanoglu and colleagues' study[5] (n = 134).**

| Associated lesion* | The present study, | | Cobanoglu, et al, | |
|---|---|---|---|---|
| | cases | % | cases | % |
| Ventricular septal defect | 17 | 51 | 71 | 53 |
| Patent ductus arteriosus | 20 | 60 | 85 | 63 |
| Atrial septal defect | 3 | 9 | 31 | 23 |
| Bicuspid aortic valve | 6 | 18 | 26 | 19 |
| Hypoplastic arch | 13 | 39 | - | |
| Aortic stenosis | 3 | 9 | 9 | 7 |
| Double-outlet right ventricle | 1 | 3 | 7 | 5 |
| Aortic regurgitation | 2 | 6 | 3 | 2 |

*One may associate with more than one lesion.

**Table 8. Management of coarctation in the present study (n = 33).**

| Treatment | Number (cases) | % |
|---|---|---|
| Medication | | |
| Anticongestive heart failure | 23 | 69.6 |
| Antihypertensive | 14 | 42.4 |
| Prostaglandin $E_1$ | 2 | 6.0 |
| Surgery | | |
| Coarctectomy with end-to-end anastomosis | 16 | 80.0 |
| Extended coarctectomy | 3 | 15.0 |
| Left subclavian flap angioplasty | 1 | 5.0 |
| Catheter intervention | | |
| Balloon angioplasty | 1 | 3.0 |
| None | | |
| Mild coarctation | 7 | 21.0 |
| Severe coarctation* | 5 | 15.0 |

* Including 1 case who denied treatment, 2 cases who lossed follow-up, and 2 cases who died preoperatively

to follow-up. There were 2 cases that died before the operation (at the age of 15 days and 1 month) and 1 case died after the operation. The overall mortality was 9 per cent with the operative mortality of 5 per cent. The causes of preoperative death were multiorgan failure due to a low-cardiac output state in one case and congestive heart failure and severe pulmonary hypertension in the other case. In postoperative death, the patient developed renal failure requiring peritoneal dialysis and pneumonia requiring long-period respiratory support since the admission. Eventually, he underwent surgery. However, chronic lung disease and cor pulmonale were developed. Sepsis was the cause of death.

The majority of the cases (21 cases, 91%) have been asymptomatic. In the hypertensive group, 9 cases underwent surgical correction with only 1 case (11%) having persistently postoperative hypertension. Mild residual CoA occurs in 1 case. A coarctectomy was performed with end-to-end anastomosis since the age of 17 days, and followed for 10 years symptom-free. The case who underwent balloon dilation of CoA had severe hypertension and has been normotensive after the procedure, asymptomatic for 3 years.

In patients associated with ventricular septal defect (VSD) where the small defect was not closed (9 cases), we found that VSD becomes smaller in 1 case (11%). However, all have been asymptomatic and free of medication.

## DISCUSSION

Coarctation of the aorta (CoA) is a common congenital heart disease in children aged 1 to 4 years. It accounts for 5-8 per cent of the patients[1] and ranks as the fourth most common congenital heart defect.[2] There is some suggestion that CoA is less common in Asian children,[1] as we found 2.1 per cent of all congenital heart defects, it was the twelfth rank in 1997[3]. Although a striking male-to-female ratio of discrete CoA, with more than two-thirds of patients being male, has been reported, this ratio is less evident in infants[1]. In the present study, 78.8 per cent of the patients presented it within the first year of life and the male-to-female ratio was 1.8:1.

### Anatomy

Adult CoA or postductal CoA is named when a short-segment abruptly narrows just beyond the insertion of the patent ductus arteriosus (PDA) or ligamentum arteriosum. The symptoms usually occur in adults. Infantile CoA, or preductal CoA is characterized by more diffuse narrowing of the aortic segment and isthmic constriction which is beyond the origin of the left subclavian artery but proximal to PDA. PDA shunts blood from the pulmonary artery into the descending Ao beyond the obstruction. It usually associates with significant congenital heart defects. Juxtaductal CoA is considered when the narrow point is opposite to the vestigial ligamentum or PDA insertion and almost always just beyond the left subclavian artery insertion, which is usually involved. A longer narrowed segment, tubular hypoplasia of the Ao, is a combination of small diameter and abnormal length, involving transverse Ao and isthmus and invariably producing obstruction. Most commonly, true tubular hypoplasia of the transverse Ao occurs in conjunction with isthmic narrowing and stenosis (true CoA)[6]. In the present study, the most common type of CoA is juxtaductal (51.5%, Table 6), compared to the Smallhorn's study in which preductal CoA is the most common type (83.3%)[4].

### Clinical manifestations

A severe-CoA infant may present, as early as the third or fourth day of life, with symptoms of advanced congestive heart failure. Sixty per cent of infants with congestive heart failure are hospitalized before the fourteenth day of life[7]. Normally, the left ventricle is under stress at birth and it may fail with the added severe CoA. This results in left atrial hypertension, persistence or resumption of fetal pulmonary hypertension, and right-to-left shunting through the PDA. The foramen ovale may be open, allowing left-to-right shunting and further aggravating the increasing pulmonary and left atrial blood flow. With the spontaneous closure of ductus arteriosus, the left ventricle assumes the added load of lower body perfusion, leading to more congestive heart failure and more pulmonary hypertension. The infant becomes acute, serious and even in low-cardiac output state. Seventy per cent of our patients presented with congestive heart failure within the first year of life. There were 2 cases manifesting with low-cardiac output state.

When VSD is associated, the usual infant with CoA becomes sick at an earlier age than does one with CoA or VSD alone. VSD carries a larger left-to-right shunt, being proportionate to the severity of CoA as well as to the size of defect[2].

According to Shinebourne's report,[8] major associated cardiac lesions were found in 85 per cent of neonates and in approximately 50 per cent of infants. In the present study, isolated CoA was found in only 12 per cent.

**Physical examination**

Comparisons of the right and left brachial and femoral pulses are important clues in the diagnosis of CoA[1,2,9]. The higher systolic blood pressures in the arm than in the leg of 20 mmHg or greater are sufficient to indicate the CoA. In the present study, we could diagnose CoA at the first examination in 85 per cent of patients by palpating the four-extremity pulses. Hypertension in the upper extremities, invariably present, may not directly reflect the severity of obstruction and may not be extreme. Thirty-six per cent of our patients had hypertension and 91 per cent of these patients were under 1 year.

Examination of the CoA patient should be systematically performed with an attempt to explain the auscultatory findings rather than with the prejudice that a particular murmur is characteristic of CoA. A murmur may be generated from the coarctation itself or collateral vessels or associated heart lesion[1].

**Electrocardiogram (ECG)**

The ECG of CoA infants is often abnormal and usually reflects coexistent anatomical defects[10]. In neonates, the ECG is usually accompanied by evidence of right ventricular hypertrophy or right bundle branch block. Left ventricular hypertrophy is apparent with more severe or long standing coarctation, though the normal ECG is often obtained in children, as seen in 33 per cent of our patients. Right bundle branch block is found in approximately 50 per cent of adults and older children and reflects a stage in progression from neonatal right ventricular hypertrophy[10].

**Chest roentgenogram (CXR)**

In infantile CoA, CXR is altered by associated defects, increased pulmonary vascular markings in left-to-right shunting, or venous congestion in severe CoA, or associated left heart obstruction. In older children, the typical CoA CXR shows normal heart size and normal pulmonary markings, unless there is an associated intracardiac shunt or left ventricular failure. The "3 sign" may be seen in AP view. Rib notching is practically pathognomonic of thoracic CoA,[11] and most commonly seen bilaterally in the 4th to 9th posterior ribs. The incidence of rib notching increases with age, uncommon below age 5, although it was visible in 14 per cent of patients between 1 and 4 years of age[12]. The majority of our patients (70%) had cardiomegaly and increased pulmonary marking (84%), reflecting the associated congenital left-to-right shunt lesion and congestive heart failure.

**Management**

Prostaglandin infusion is particularly applicable to the sick baby with severe heart failure or shock[13,14]. Congestive heart failure is further improved by furosemide and inotropics. The goal of medical treatment is to stabilize the patient preoperatively for surgery. Treatment of hypertension by medical means alone is usually impossible and causes danger[1]. Surgical therapy remains the definite means of effective relief of obstruction. The hospital mortality in the current era is low (2% to 10%) in neonates undergoing an operation for isolated coarctation and hypoplastic left heart class I (uncomplicated coarctation), with or without PDA. When coarctation is repaired in older infants, children, adolescents and young adults, the early mortality is about 1 per cent.[15] The optimal time for surgery is considered when the patient is aged 5 to 7[1]. However, most pediatric cardiologists recommend elective correction of CoA for all patients beyond the age of 3 years but before age 10, despite a lack of symptoms or severe upper extremity hypertension. It has been documented that the diameter of aorta at 3 years of age is 55 per cent of the diameter of adulthood[16]. An uncomplicated CoA with a consistent systolic blood pressure gradient between the arms and legs of more than 20 mmHg is an indication for surgical correction[2].

Three preferred techniques are 1) resection and end-to-end anastomosis; 2) subclavian flap aortoplasty, and 3) synthetic patch aortoplasty,[17] as in our institute. The incidence of recurrent coarctation in CoA patients who underwent coarctectomy under the age of 3 months was lower in an end-to-end anastomosis operation compared to subclavian flap aortoplasty (5-year surgery free 92±5% *vs* 75±7%, respectively). However, operative mortality rate was not different[5]. The overall mortality in

the present study was 9 per cent, and the operative mortality was 5 per cent. All patients who died were under 1 year of age.

Percutaneous transluminal angioplasty was demonstrated to be effective in patients who developed restenosis after the surgery[18-20]. Aneurysm formation does not invariably occur, however, it is rare on short-term prospective follow-ups in some institutions[21]. The use of dilation in infants is not clearly established. Histologic studies of patients with large areas of ductal tissue in the area of constriction project that balloon angioplasty will palliate only temporarily, and that restenosis is inevitable[22,23].

**Natural history and complications**

CoA was accompanied by a 90 per cent mortality by age 50. The mean age of death was 35 years. The causes of death were intracranial hemorrhage (11% of cases in autopsy series), aortic rupture or dissection (23%), endocarditis (22%), and congestive heart failure (18%)[24-26]. Prolonged preoperative hypertension and surgery after the age of 25 are associated with a risk of premature cardiovascular death[1]. The degree of preoperative hypertension has some influence on whether hypertension, thus, will recur or be poorly relieved by operation[27-29]. Liberthson, and colleagues[30] followed 234 patients, 40-years postoperation, and found that the incidence of persistently postoperative hypertension in patients performing surgery at the age of 1-5 years was 6 per cent, at the age of 6-18 years was 30 per cent, and at the age of 18 years up was 47 per cent. Aortic aneurysm is a long-term sequelae of repairs using patch graft and tubular graft techniques[31]. Residual gradients and the need for reoperation are most often found when surgery is necessitated in a young patient of less than 2 months[2]. The age, seemingly, has greater influence than the technique used[32]. In one study, the reintervention rate for residual or recurrent coarctation was 9 per cent[17]. In the present study, we had only 1 case who has mild residual CoA and no intervention is needed. He underwent surgery since the age of 17 days. However, long-term follow-up is required.

## SUMMARY

Coarctation of the aorta is a common congenital heart disease. Comparison of the pulse and blood pressure between upper and lower extremities is an important clue for the diagnosis. Early diagnosis and proper intervention can reduce mortality and morbidity, especially in a sick baby with severe congestive heart failure and low cardiac output. The effective means to relieve the obstruction is surgical repair. However, percutaneous transluminal angioplasty has been demonstrated to be effective in patients who develop restenosis after the operation. Long-term follow-up is required to prevent complication, whether or not surgery is performed.

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# การตีบแคบของหลอดเลือดแดงเอออร์ต้าในผู้ป่วยเด็กที่โรงพยาบาลศิริราช

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ได้ทำการศึกษาย้อนหลังในผู้ป่วยเด็ก จำนวน 33 ราย ที่ได้รับการวินิจฉัยว่ามีการตีบแคบของหลอดเลือดแดงเอออร์ต้า จากภาควิชากุมารเวชศาสตร์ โรงพยาบาลศิริราช ตั้งแต่เดือนมกราคม 2532 ถึง ธันวาคม 2541 พบว่าเป็นเพศชาย 21 ราย (ร้อยละ 64) และเพศหญิง 12 ราย (ร้อยละ 36) อายุตั้งแต่ 1 วัน ถึง 11 ปี (ค่ามัธยฐาน 2 เดือน) ส่วนใหญ่ของผู้ป่วย (ร้อยละ 78.8) ได้รับการวินิจฉัยภายในอายุขวบปีแรก อาการและอาการแสดงที่สำคัญ ได้แก่ หัวใจวาย (ร้อยละ 69.6) ความดันโลหิตที่แขนสูง (ร้อยละ 36.3) และชีพจรที่ขาเบากว่าแขน ภาพถ่ายรังสีทรวงอก พบเงาหัวใจโต (ร้อยละ 70) และปริมาณหลอดเลือดไปปอดเพิ่มขึ้น (ร้อยละ 84) ซึ่งสัมพันธ์กับภาวะหัวใจวายและโรคหัวใจพิการแต่กำเนิดชนิด left-to-right shunt ที่พบร่วม ผลการตรวจคลื่นไฟฟ้าหัวใจ พบว่าปกติ (ร้อยละ 33.3) เวนตริเคิลขวาโต (ร้อยละ 33.3) เวนตริเคิลซ้ายโต (ร้อยละ 22.2) และเวนตริเคิลทั้งสองข้างโต (ร้อยละ 11.2) ในผู้ป่วยที่อายุน้อยจะพบว่าเวนตริเคิลขวาโตบ่อยกว่า จากการตรวจหัวใจด้วยคลื่นเสียงสะท้อนความถี่สูง และ/หรือการฉีดสารทึบรังสีบริเวณหลอดเลือดแดงเอออร์ต้า พบรอยตีบแคบชนิด juxtaductal บ่อยที่สุด (ร้อยละ 51.5) รองลงมาคือ postductal (ร้อยละ 27.3) และ preductal (ร้อยละ 21.2) การรักษาทางยาประกอบด้วย การควบคุมภาวะ low cardiac output ด้วย prostaglandin $E_1$ ในทารกแรกเกิด การควบคุมภาวะหัวใจวาย ความดันโลหิตสูง และการทำผ่าตัดแก้ไขภาวะตีบแคบของหลอดเลือดในรายที่มีข้อบ่งชี้ พบว่าได้ผลดี และมีอัตราเสี่ยงต่ำ (ร้อยละ 5) อัตราตายในการศึกษาครั้งนี้ ร้อยละ 9 สาเหตุการตายที่สำคัญคือ อวัยวะล้มเหลวจากภาวะ low cardiac output และการติดเชื้อในปอด อัตราการเกิดการตีบแคบหลงเหลือหลังการผ่าตัดพบร้อยละ 5

**คำสำคัญ** : การตีบแคบของหลอดเลือดแดงเอออร์ต้า, ผู้ป่วยเด็ก

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* ภาควิชากุมารเวชศาสตร์, คณะแพทยศาสตร์ศิริราชพยาบาล, มหาวิทยาลัยมหิดล, กรุงเทพฯ 10700