

# Truncus Arteriosus: Siriraj Experience

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## ABSTRACT

Between January 1<sup>st</sup>, 1995 and December 31<sup>st</sup>, 2004, a total of 30 children (17 girls, 56.7% and 13 boys, 43.3%) were diagnosed to have truncus arteriosus at the Department of Pediatrics, Siriraj Hospital. The ages at the first diagnosis ranged from 1 day to 3 years (median 120 days). The predominant clinical presentations were congestive heart failure (53.3%), cyanosis (30%) and feeding problems (26.7%). All patients had heart murmur. Chest roentgenogram demonstrated cardiomegaly and increased pulmonary vascularity in 86.7% and 83.3%, respectively. Electrocardiogram showed a frontal plane QRS axis in a range of 0-90° in 70% of the patients, left ventricular hypertrophy and biventricular hypertrophy in 50% and 40%, respectively. Echocardiogram revealed type I anomaly in the majority of the patients (80%), and type II in the rest of patients. Ten percent of the patients had right-sided aortic arch. The majority of the patients received more than one medication for controlling heart failure. Sixteen patients (53.3%) were operated at the median age of 133 days and median weight of 3.8 kilograms. Thirteen percent of the patients (2 cases) had palliative surgery (pulmonary artery banding) and 87.5% of patients (14 cases) underwent total correction. All patients who had total repair had immediate complications, of which the majority (57.1%) were pulmonary hypertensive crisis. There were a total of 9 deaths (30%); 7 patients died immediately (<14 days) post total repair, 2 patients died preoperatively. During follow-up (median 57.9 months), all patients were asymptomatic except one patient (90.9%) who was re-operated on for conduit replacement due to severe stenosis and truncal valve repair due to severe regurgitation at 22 months after the first operation. Three patients have been waiting for surgery. In the present study, we could not definitely relate the associated risk factor with mortality such as the age at surgery, preoperatively high pulmonary vascular resistance or truncal valve abnormality. However, it seems that the older age at surgery may be the possible risk factor. Therefore, early diagnosis and surgical intervention for this anomaly should be considered.

**Keywords:** Common arterial trunk; truncus arteriosus

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**T**runcus arteriosus is an uncommon congenital cardiovascular malformation, classified as a conotruncal defect or defect of the outflow tract of the heart. Overall, the reported prevalence ranges from 0.04-0.09 cases per 1,000 live births and accounts for 1.1-2.5% of all types of congenital heart disease.<sup>1-3</sup> It usually occurs as an isolated cardiovascular malformation, although on occasion it has been reported to be associated with anomalies of other organ systems, particularly the DiGeorge's syndrome.<sup>4</sup> Thirty four percent of all patients with truncus arteriosus had a 22q11 deletion.<sup>5</sup> An infant of a diabetic mother has approximately a 12-to 13-fold increased risk of developing truncus arteriosus compared with the infant of a non-diabetic mother.<sup>6</sup> In Thailand, It has been reported that truncus arteriosus was found in 1.3-1.8% of congenital

heart diseases under one year of age.<sup>7,8</sup> However, the details of this malformation and results of the treatment have not been mentioned.

The natural survival of this condition is usually short. The mean age of death was 5 weeks. Survival beyond the age of 1 year was only 15%.<sup>3</sup> This dismal natural history was the main factor that gave rise to the approach of early surgical intervention advocated for these patients. The purpose of this study was to analyse the demographic data, clinical presentations, investigation and the outcome of these patients in our 10-year experience.

## MATERIALS AND METHODS

All patients were diagnosed as having truncus arteriosus by echocardiogram at the Department of Pediatrics, Faculty of Medicine, Siriraj Hospital between January 1995 and December 2004. Demographic data, clinical manifestations,

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and investigations including chest roengenographic, electrocardiographic, echocardiographic and cardiac catheterization reports were retrospectively reviewed from the medical records and echocardiographic and the cardiac catheterization database.

## RESULTS

Between January 1<sup>st</sup>, 1995 and December 31<sup>st</sup>, 2004, there were 30 cases of truncus arteriosus diagnosed at our institution, comprising of 17 females (56.7%) and 13 males (43.3%). The sex ratio of female: male was 1.3:1. The median age at the first diagnosis at our hospital was 120 days (range 1 day to 3 years, Table 1).

### Clinical manifestations

The most common clinical presentation in the present study was congestive heart failure (53.3%, Table 2).

### Physical examination at the first diagnosis

Tachypnea was found in 76.7%, hepatomegaly in 86.7%. Cardiac murmur was heard in every case. Types of ausculted cardiac murmurs in the present study were systolic murmur (22 cases, 73.3%), both systolic and diastolic (to and fro) murmur (6 cases, 20%) and continuous murmur (2 cases, 6.7%). Transcutaneous oxygen saturations in the majority of our patients (46.7%) were in the ranges of 80-89%, in 23.3% they were between 90-94% and in 20.2% they were > 95%. The associated extra-cardiac anomalies were found in 7 cases (23.3%); there were DiGeorge syndrome, congenital iris cyst, delayed development, Jacobson syndrome (11q23 deletion), microcephaly, micrognathia, and congenital tracheal and subglottic stenosis, one of each.

### Investigation

The average of hematocrit level in our patients was 41 + 7.3%. Hematocrit level less than 40% was found in 53.3% (16 cases), in the range of 40-49% in 23.3% (7 cases), in the range of 50-59% in 6.7% (2 cases) and > 60% in 3.3% (1 case). Chest roentgenogram in the majority of our cases (26 cases, 86.7%) demonstrated cardiomegaly by increasing cardiothoracic ratios ranging from 0.52 to 0.73 (mean 0.6 + 0.04). Twenty-five cases (83.3%) showed increased pulmonary blood flow. Electrocardiogram revealed axis 0-90° in 21 cases (70%), and 90-180° in 9 cases (30%). Left ventricular hypertrophy was found in 15 cases (50%), biventricular hypertrophy in 12 cases (40%), right ventricular hypertrophy in 3 cases (10%) and left atrial enlargement in 4 cases (13.3%).

Echocardiogram demonstrated type I anomaly, classified by Collett and Edwards in 24 cases (80%) and type II in 6 cases (20%). Truncal valve abnormalities were found in 18 cases (60%); including 16 cases (88.9%) of insufficiency (mild degree in 30%, moderate degree in 20% and severe degree in 3.3% of the patients) and 2 cases of stenosis (11.1%). Three cases (10%) had right-sided aortic arch

**TABLE 1.** The patients' age at the first diagnosis at our hospital (n = 30)

Age at diagnosis	Cases (%)
Newly born - 1 month	8 (26.7)
1 month - 1 year	16 (53.3)
> 1 year	5 (16.7)
No data available	1 (3.3)
<b>Total</b>	<b>30 (100.0)</b>

**TABLE 2.** Clinical manifestations in the present study (n = 30)

Symptoms & signs	Cases (%)
Heart failure	16 (53.3)
Cyanosis	9 (30.0)
Feeding problem	8 (26.7)
Asymptomatic heart murmur	8 (26.7)
Failure to thrive	3 (10.0)
Tachypnea	4 (13.3)
Bleeding disorder*	1 (3.3)

Some cases had more than one clinical manifestation

\* associated with Jacobson syndrome

and 1 case had bilateral superior vena cava.

Preoperative cardiac catheterization and angiogram were performed in 11 cases (36.7%). In all cases, the findings were similar to the echocardiographic reports, except one in whom the cardiac catheterization report was type I but the echocardiographic report was type II. The pulmonary vascular resistance (PVR) was ranged between 2.7-34 Wood unit. m<sup>2</sup> (median 8 Wood unit. m<sup>2</sup>) in room air. Five cases (41.7%) underwent total correction, and 2 cases of them died within 14 days postoperatively due to a pulmonary hypertensive crisis (preoperative PVR 3.3 and 8 Wood unit. m<sup>2</sup>).

### Management and outcome

The majority of our patients received more than 1 class of anticongestive heart failure medications. Management and outcome of these patients were summarized in Diagram 1. Cardiac surgery was performed in 16 cases (53.3%); palliative surgery (pulmonary artery banding 2 cases, 12.5%) at the ages of 2 and 5 months, one because of bleeding tendency (Jacobson syndrome), and total correction (patch closure ventricular septal defect and right ventricular to pulmonary artery homograft placement 14 cases, 87.5%) at the ages ranging from 47 to 332 days (median 133 days), with body weights ranging from 3.2 to 6.3 kg. (median 3.8 kg).

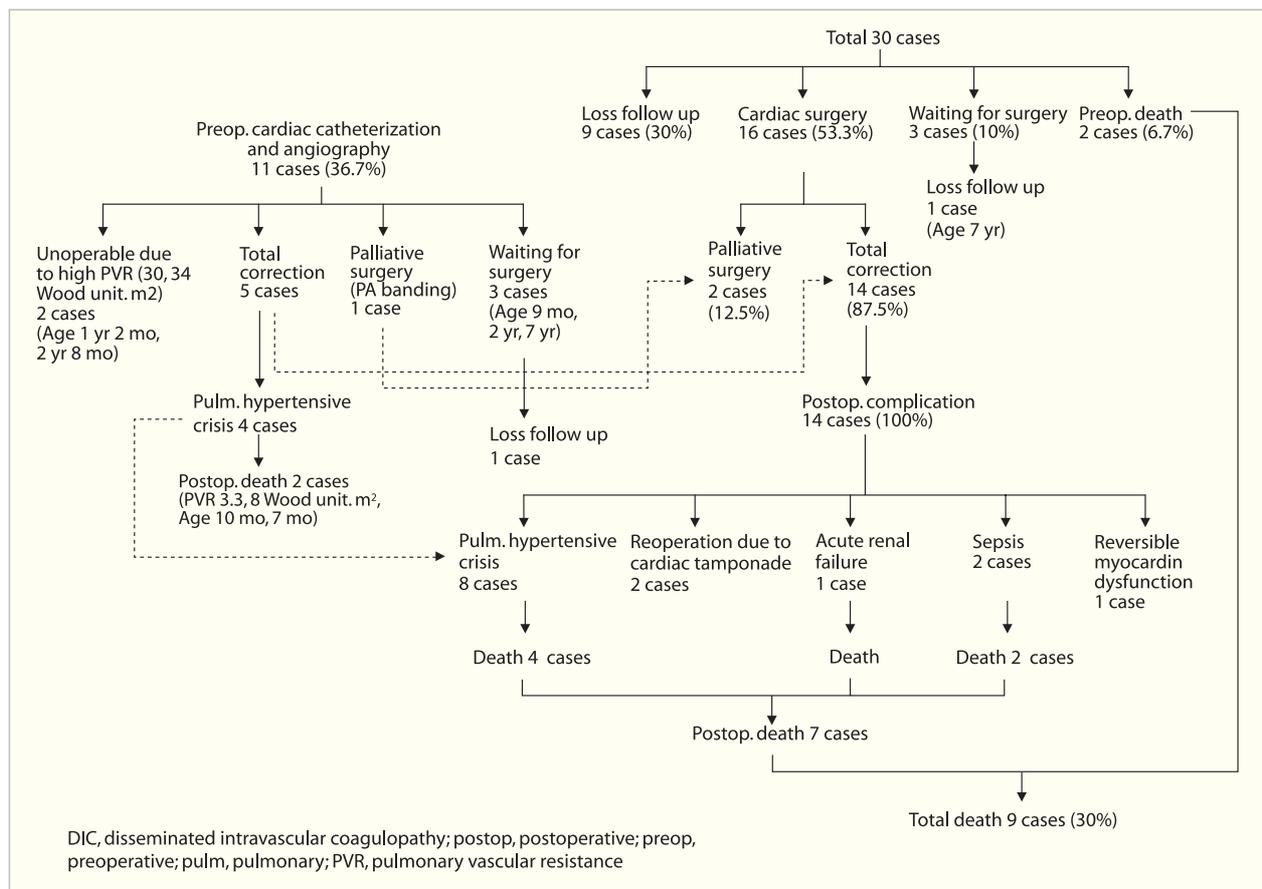
Pulmonary hypertensive crisis was the most common immediate complication (8 cases, 57.1%) within 7 days after the total correction. Nine cases (30%) died at the age of 4-12 months ; 7 cases died within 14 days after total correction with the median age of 7 months and 2 cases died preoperatively (aged 15 days and 7 months) due to sepsis and disseminated intravascular coagulation. The mortality rate of the disease was 30% and the mortality rate of the total repair was 50%. We could not definitely relate the risk factors with mortality such as the age at surgery, preoperatively high pulmonary vascular resistance or truncal valve abnormalities due to having few cases in each group.

At the most recent follow-up period, ranging 2.1-95 months (median 57.9 months), 11 patients were regularly followed up and 10 cases were asymptomatic; 4 cases were post total correction, 1 case was post pulmonary artery banding, 3 cases were waiting for surgery and 2 cases were inoperable, due to high risk (aged at the first visit at our hospital of 1 year and 2 months and 2 years and 8 months). One case was re-operated after 22 months of surgery for changing the conduit and repairing of severe regurgitation truncal (aortic) valve, without any complication.

## DISCUSSION

Truncus arteriosus is an uncommon congenital heart defect. Gender difference in frequency is not striking,

**DIAGRAM 1.** Management and outcome in the present study



although most series reported more males than females.<sup>3</sup> In the present study, the female: male ratio was 1.3:1. The majority of patients are recognized during early infancy, often during the neonatal period.<sup>3</sup> The median age at the first diagnosis in the present study was 120 days, most likely due to the fact that most patients were referred to us from other hospitals. Eighty percent of the cases presented within the first year of life. Congestive heart failure was the most common clinical presentation in our patients (53.3%), secondary to the increased volume load on the heart produced by the excessive blood flow through the pulmonary circulation. Truncal valve insufficiency, moderate and severe degrees were found in 20% and 3.3% of cases respectively in the present study, which resulted in additional volume load on the heart and early heart failure. Cyanosis was found in 30% of the cases. During the first week of life, persistent elevation of pulmonary vascular resistance can cause mild cyanosis. Stenosis of the pulmonary arteries may be the other uncommon cause of intensifying cyanosis with increasing age. A loud pansystolic murmur maximally heard at the lower left sternal border with radiation to the entire precordium was the most frequent finding in the present study (73.3%). Left ventricular hypertrophy demonstrated by electrocardiogram was seen predominantly in patients with large pulmonary blood flow, as shown in our patients (50%). Patients with normal or decreased pulmonary flow may exhibit right ventricular hypertrophy only. Typically, chest roentgenogram shows moderate cardiomegaly with increased pulmonary vascular markings. The right-sided aortic arch is found in approximately 21-36% of the patients,<sup>10,11</sup> and 10% of the present study. The combinations of right aortic arch and increased pulmonary vascularity in mild cyanotic patient are strongly suggestive

of truncus arteriosus. The disproportionate enlargement of the central pulmonary arteries associated with accentuated tapering of the distal pulmonary arterial tree reflects pulmonary vascular obstructive disease, which is commonly developed in the early age.

Two-dimensional and Doppler echocardiography including color-flow technique has greatly increased the ability to determine accurately the cardiac anatomy and, in some cases, the hemodynamics in truncus arteriosus, even in utero.<sup>9</sup> Cardiac catheterization and angiography are now infrequently necessary in the patients with truncus arteriosus, except for assessing the status of the pulmonary vascular bed. Delay of operation results in chronic ischemia of the hypertrophied ventricle and pulmonary vascular obstructive disease, which related to the mortality of operation and disease. The choice of operation is between total correction and pulmonary artery banding which is now reserved for the few patients in whom the risk of total repair is prohibitive because of associated significant cardiovascular malformations and/or other conditions such as sepsis or shock or bleeding disorder as one of our patients (Jacobson syndrome). Corrective surgery at 6-12 months of age is associated with a mortality that is twice the repair between 6 weeks and 6 months of age.<sup>13</sup> The operative mortality rate of total correction in patients without severe abnormalities was 5-30%.<sup>12,14</sup> In the present study, the operative mortality rate was 50%. The most common causes of death were chronic heart failure and high PVR resulting in postoperative pulmonary hypertensive crisis and complicated preoperative infection. Although, in this era, many pulmonary vasodilating therapies are effective, there is a high morbidity and mortality from postoperative pulmonary hypertensive crisis which relates to the older

age at surgery which still occurs. Overall hospital mortality was 25%,<sup>12</sup> and 30% in the present study. Because the truncal valve is often abnormal, the potential exists even after corrective operation for the gradual progression of truncal (aortic) valve incompetence eventually requiring valve replacement. In patients assessed preoperatively to have mild, moderate, or severe truncal valve incompetence, the eventual probability of the need for aortic valve replacement was high. This is in contrast to the patients in whom the truncal valve incompetence was assessed as none or minimal at the time of correction, none of these patients required subsequent aortic valve replacement.<sup>3</sup> In patients who undergo successful correction during early infancy, the small conduit eventually must be replaced with a larger one in 28.5% of the cases, however re-operation for conduit replacement alone carries a low risk.<sup>15</sup>

Overall actuarial survival to 24 years was 46.4%. Late survival in patients judged to have no or mild aortic truncal valve incompetence at the time of corrective operation was 57% at 25 years, compared to 21% in patients in whom the aortic valve was judged to be moderately or severely incompetent.<sup>3</sup> Late deaths which did not occur at the time of re-operation, most often were caused by progression of pulmonary vascular disease, continued progressive left ventricular deterioration and congestive heart failure, bacterial endocarditis and sudden death (presumed dysrhythmia). However, there is a much improved long-term outcome for patients undergoing correction in the first 2 years of life compared with those undergoing correction beyond 2 years of age which was reported.<sup>3</sup> In our experience, delay in operation will increase the risk of infection; especially pneumonia, chronic congestive heart failure and high operative mortality and morbidity.

## SUMMARY

The prognosis of truncus arteriosus is generally poor. Early surgical intervention is now advocated for these patients at around the age of 6 weeks. Delay in operation will increase risk of preoperative infection and high pulmonary vascular resistance which relates to postoperative pulmonary hypertensive crisis and high mortality and morbidity. Long term follow-up, even in successfully

operated patients, to monitor for progressive pulmonary vascular disease, truncal (aortic) valve incompetence and/or stenosis, (which may require re-operation for homograft replacement) and in infective endocarditis, it is mandatory.

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