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## ORIGINAL ARTICLE

# Some Aspects of Tricuspid Atresia

*by*

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**Abstract**

*During the period of 5 years, from the beginning of 1971 up to the end of 1975, out of 2054 sick children seen in the Pediatric Outpatient Division of the Department of Cardiology Medical School, University of Indonesia/Dr. Cipto Mangunkusumo General Hospital Jakarta, 1073 patients were found to have congenital heart disease.*

*Ten out of these 1073 CHD patients have been diagnosed as Tricuspid Atresia on the clinical basis, giving the incidence of 0,93% of all CHD, of whom 5 were boys and 5 were girls, mean age being 36,5% months (range 2 months to 12 years).*

*Hemodynamic studies have been done to 3 of the 10 patients, all of which confirmed the diagnosis. Two children went to surgery.*

*Anatomic and radiologic classification and types of the disease in those 3 children and some clinical aspects and hemodynamic studies are discussed.*

*It is stressed that early recognition and surgical intervention are indicated, and still should be improved.*

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## Introduction

Tricuspid Atresia as an anatomical entity was recognized as early as 1823 by Holmes, as cited by Dick et al. (1975). Since then many publications of this disease as a clinical entity, have been described in textbooks as well as in various medical journals.

It may be defined as a congenital complete absence of the right sided atrioventricular valve, with hypoplasia of the right ventricle, usually associated with patency of the interatrial septum and hyperplasia of the mitral valve and left ventricle. (Kjellberg et al. 1955; Keith and Vlad 1967; Nadas and Fyler 1972). Variations of clinical features have made many authors to differentiate various anatomical classifications of this disease.

Edward and Burchell (1949) introduced 2 groups, with and without transposition of the great vessels, and they described 2 types in each group. Keith and Vlad (1967) amplified this anatomical classification to include 8 types in all: 3 types in the 1st group without transposition, (a) with Pulmonary Atresia and intact ventricular septum, (b) with Pulmonary hypoplasia and small ventricular septal defect, (c) with normal Pulmonary artery and large ventricular septal defect; 3 types in the 2nd group with d- transposition, (a) with Pulmonary Atresia, (b) with Pulmonary/Subpulmonary stenosis, (c) with large Pulmonary artery, and 2 types in the 3rd group with l- transposition, (a) with Pulmonary/Subpulmonary stenosis, (b) with Sub-aortic stenosis. Recently Dick et al. (1975) in the review of their 101 cases added 3 groups of radiological classification to each of the 3 types of anatomical classification described by many authors before so that they had 9 types in all. Radiological groups they mentioned are: group-A with decreased lung vascularity, group-B with normal or increased lung vascularity, and group-C with changing lung vascularity.

The prevalence of this disease accounts for approximately 1 to 3% of all congenital heart defects in various centers (Friedberg, 1972; Hanafiah et al., 1965; Maemunah S.A. et al., 1974; Kaplan, 1969; Nadas et al., 1972). The most frequent type reported in literature is of the type I anatomy (70%), while group-A was said to be the most common radiological group (62%).

Prognosis of Tricuspid Atresia was said to be very poor. It was generally considered as an anatomically uncorrectable lesion.

In a review of the age of death in 111 cases Keith and Vlad (1967) disclosed that 49.5% died before the age of six months; 66% at one year of age, while only 10% survived over the age of 10 years. A later report by Dick et al. (1975) mentioned that their patients of the most frequent category, type I group A, without surgery had only 10% chance of survival to 15 years while sur-

gical intervention improved chance of survival to 15 years of age to 50%. They also concluded that the continued need of early surgical palliation for this disease are indicated.

Anastomosis of the right auricular appendage to the pulmonary artery as described by Hurwitt et al. (1955), has been developed to right atrium-pulmonary artery allograft by Fontan and Baudet (1971). Stanford et al. (1973) followed the later procedure and also described it as a corrective surgical repair for Tricuspid Atresia. Many experts are of the opinion that this new surgical approach may result in a more prolonged survival.

It is the purpose of this study to learn and to present some aspects of Tricuspid Atresia in our clinic in order to ask more attention and to increase awareness to this clinical entity, in connection with the hopeful reports that early recognition and surgical intervention may greatly improve its prognosis.

## Materials and method

This study has been done retrospectively. Medical records of patients in The Pediatric Outpatient Division of the Department of Cardiology in the period of 1971 — 1975; and those of patients in the Inpatient Division, in the Hemodynamic Division and in the Surgical Division of The Department of Cardiology in the same period were selected and reviewed. Hemodynamic studies were performed by two of the authors.

### *Criteria for diagnosis*

Many authors have described in various textbooks and other publications that Tricuspid Atresia should be highly suspected on the basis of clinical symptoms and signs, radiological and electrocardiographic abnormalities. Hemodynamic studies with cardiac catheterization and angiocardiography may confirm the diagnosis and show various anatomical types. Direct sight during surgery will certainly also give more information. At last, autopsy will verify the precise diagnosis, including the anatomical types of the lesion, through macroscopic and microscopic examination.

### *Clinical*

Symptoms and signs of early cyanosis, central and progressive, with or without murmurs, clubbing, frequent episodes of hypoxic spell, growth retardation, fatigue, squatting, and other symptoms and signs of cyanotic Congenital Heart Disease or sometimes symptoms and signs of congestive heart failure.

### *Radiological abnormalities.*

The cardiac contour shows right heart border or prominent right atrial border, slight to moderate cardiomegaly, blunt apex with a concavity or lack of prominence of Pulmonary Artery area. The pulmonary vasculature is usually decreased (75%), except when associated with Transposition of the Great Arteries

or large Ventricular Septal Defect. (Elliot and Schieber 1971; Franch, 1969 and Nadas and Fyler 1972).

*Electrocardiographic abnormalities.*

Many authors (Taussig, 1960; Perloff, 1970; Sodi Pallares, 1970; Elliot, 1971; Friedman, 1971 and Marriot, 1975) described that some ECG abnormalities are very specific for Tricuspid Atresia, those are : Left Axis Deviation, horizontal electrical position, Left Ventricular Hypertrophy, Right Atrial Hypertrophy (P-congenitale), or Biatrial Hypertrophy with the right side predominant (P-tricuspidale).

*Hemodynamic studies.*

Kjellberg (1955), Keith et al., (1967), Diehl et al. (1968) and Nadas and Fyler (1972) described that on catherization, catheter cannot go directly from the Right Ventricle. The route of catheter is from the Right Atrium to the Left Atrium and then to the Left Venricle. The Right Atrial Pressure, either a wave or mean pressure, exceeds those of Left Atrium by varying degrees. Systemic 02 saturation from Left Atrium downward is lower than normal.

Angiocardiography from Right Atrium will show some special features:

a) typical sequence of opacification from the large Right Atrium to the Left Atrium, to the Left Ventricle, and then to the Great Arteries;

b) specific non-opacification of the inflow tract of the Right Ventricle;

c) radioluscent triangular Right Ventricular window between the medial edge of the shadow cast by the prominent Inferior Caval Vein and the shadow of the Left Ventricle on the other. Baron (1971) mentioned this sign as pathognomonic for Tricuspid Atresia. Selective angiocardiography from the Left Ventricle will show anatomical classification. Keith and Vlad (1967) in their series of 143 cases mentioned the figures as follows :

| | | | | | |
|---|---|---|---|---|---|
| I | a. 13<br>b. 73<br>c. 13 | 69% | II | a. 3<br>b. 11<br>c. 26 | 28% |
| III | a. 1<br>b. 3 | 3% | | | |

*Management*

A. Medical : especially for symptomatic purposes.

B. Surgical :

1. Palliative surgery :
   a. To increase Pulmonary blood flow when reduced : with arterial shunting (Waterston/Potts/Blalock Taussig procedure), or with venous shunting (Glenn procedure).
   b. To remove intracardiac barrier which interfere with free flow of blood (Rashkind/Blalock Hanlon procedure).
   c. To diminish Pulmonary blood flow when excessive (Main Pulmonary Artery Banding).

2. Corrective (?) surgery :

Fontan Baudet procedure which consists of : (a) Aortic allograft conduit is inserted between Right Atrium and Left Pulmonary Artery, (b) Pulmonary valve allograft is sutured at the Right Atrium — Inferior Vena Caval orifice, (c) Superior Vena Caval — Right Pulmonary Artery shunt (Glenn) is also carried out, (d) closure of Atrial Septal Defect if present.

## Results

From 2054 sick children seen in the Pediatric Out-Patient Division of the Department of Cardiology in that period, 1073 patients were found to have congenital heart disease (CHD).

Ten out of these 1073 CHD patients have been diagnosed as Tricuspid Atresia on the clinical basis (0.93%), of whom were 5 boys and 5 girls, mean age 36.5 months (range 2 months — 12 years).

Only 3 of the 10 patients underwent further hemodynamic studies, all of which confirmed the diagnosis, and 2 of these 3 had surgical intervention. In this connection for further detailed description we prefer to limit the discussion to only 3 catheterized children : (1) Sa, female, 5 years; (2) Su, male, 6 months; (3) J.K., male, 12 years. See tables 1-2 and figures 1-3 below.

TABLE 1 : *Clinical findings*

| | Sa, ♀, 5 | Su, ♂, $\frac{6}{12}$ | J.K. ♂, 12 |
|---|---|---|---|
| HISTORY : | | | |
| antenatal | normal | normal | normal |
| natal | normal | normal | normal |
| early progressive central cyanosis | ++ | ++ | + |
| increasing dyspnoea | ++ | ++ | + |
| feeding difficulty | ++ | ++ | + |
| hypoxic spells | ++ | ++ | + |
| squatting | ++ | ++ | + |
| growth retardation | ++ | ++ | mild |
| EXAMINATION : | | | |
| undernourished | ++ | ++ | mild |
| cyanosis | ++ | ++ | mild |
| clubbing | ++ | + | + |
| bulging of the chest | — | — | — |
| RV impulse | — | — | — |
| second heart sound ($S_2$) | single | single | single |
| pulm. ej. syst. murmur | harsh | harsh | harsh |
| heart failure | — | — | — |

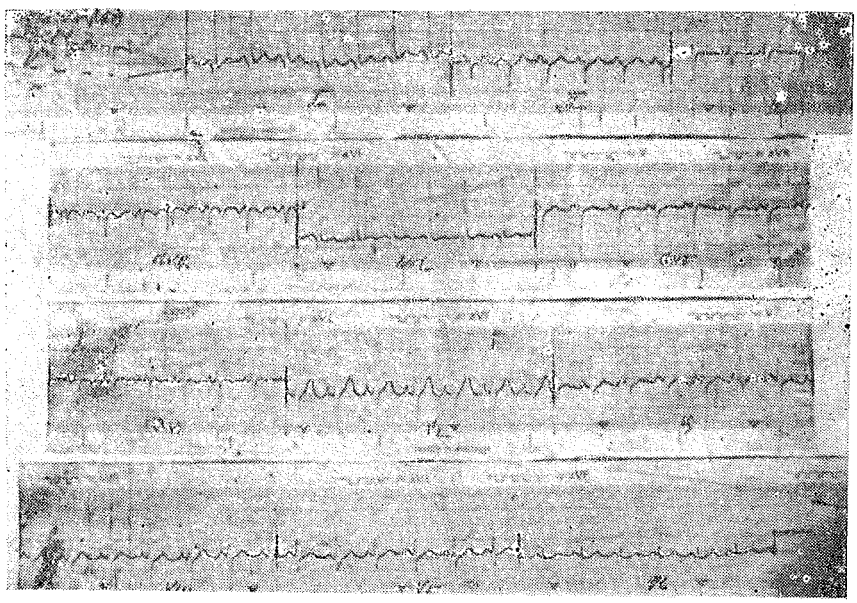
TABLE 1: *Clinical findings (continued)*

| | Sa, ♀, 5 | Su, ♂, $\frac{6}{12}$ | J.K., ♂, 12 |
|---|---|---|---|
| Chest X Ray (see also fig. 1 - 3) | sl. cardiomegaly<br>left Aortic arch<br>widened Aorta<br>decr. lung vasc.<br>— | sl. cardiomegaly<br>left Aortic arch<br>widened Aorta<br>straight right heart border | sl. cardiomegaly<br>left Aortic arch<br>widened Aorta<br>decr. lung vasc.<br>— |
| ECG (see also fig. 1, 2, 3). | LAD -40°<br>P tricuspidale<br>LVH, $qV_5V_6$ absent | LAD -15°<br>P tricuspidale<br>LVH, $qV_5V_6$ absent | LAD -45°<br>P tricuspidale<br>LVH, $qV_5V_6$ absent |
| Laboratory | Hb 18 gm%<br>Ery 6.82 mill<br>Ht 78% | Hb 17.5 gm%<br>Ery 6.35 mill<br>Ht 62% | Hb 17,5 gm%<br>Ery 5.68 mill<br>Ht 55% |
| Neurology | EEG abnormal (irritative) | EEG dubious record | EEG abnormal (slowering activity) |
| Ophthalmology | retinopathy grade I<br>ec. polycythemia | not examined | retinopathy grade I<br>ec. polycythemia |

TABLE 2 : *Hemodynamic studies*

| | Sa, ♀, 5 | Su, ♂, $\frac{6}{12}$ | J.K., ♂, 12 |
|---|---|---|---|
| CATHETERIZATION : | | | |
| route of catheter | RA → LA → LV | RA → LA → LV | RA → LA → LV |
| pressure | RA > LA | RA > LA | RA > LA |
| $0_2$ saturation : | | | |
| RA | 41% | 56% | 70% |
| LA | 45% | 64% | 73% |
| LPV | 100% | 95% | 96% |
| LV | 50% | 62% | 72% |
| ANGIOCARDIOGRAPHY: | | | |
| typical sequence | RA ⇒ LA → LV → Aorta | RA ⇒ LA → LV → Aorta | RA ⇒ LA → LV → Aorta |
| RV window | present | present | present |
| Pulmonary Artery filling | poor | poor retrogradely | poor |

LAD = Left Axis Deviation
LVH = Left Ventricular Hypertrophy
RA = Right Atrium
LA = Left Atrium
LV = Left Ventricle
LPV = Left Pulmonary Vein
sl. cardiomegaly = slight cardiomegaly
Ht = Hematocrit
EEG = Electroencepalography
age : 5 = 5 years
$\frac{6}{12}$ = 6 months
12 = 12 years.



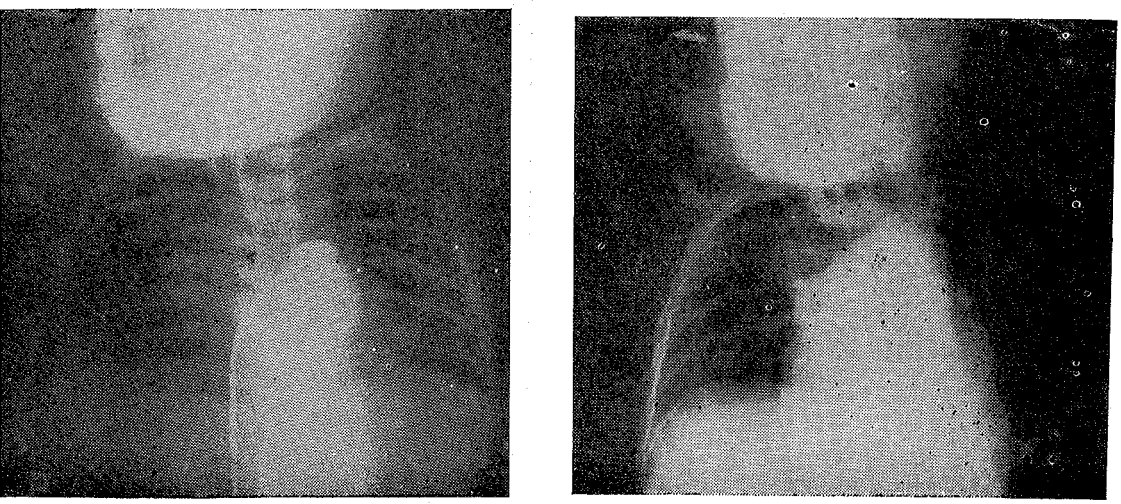
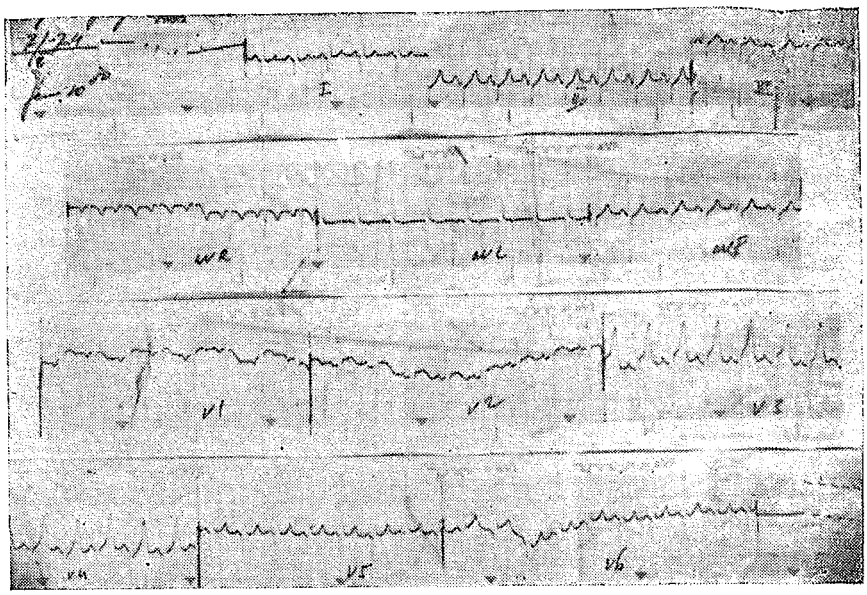
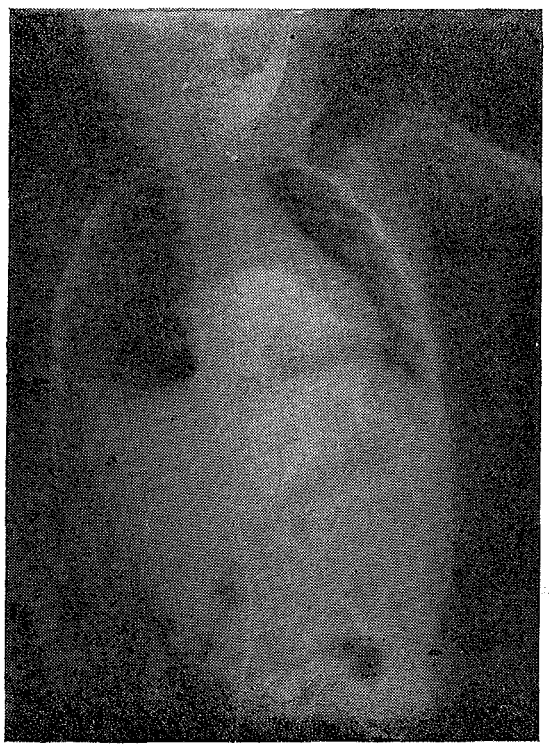


FIG. 1 : *ECG and 2 pieces of angiocardiographic pictures of case 1 : Sa, ♀, 5 years*





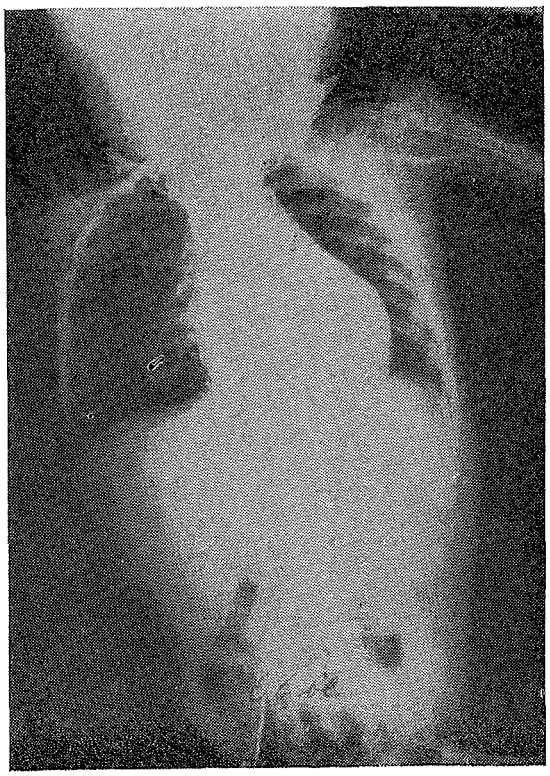
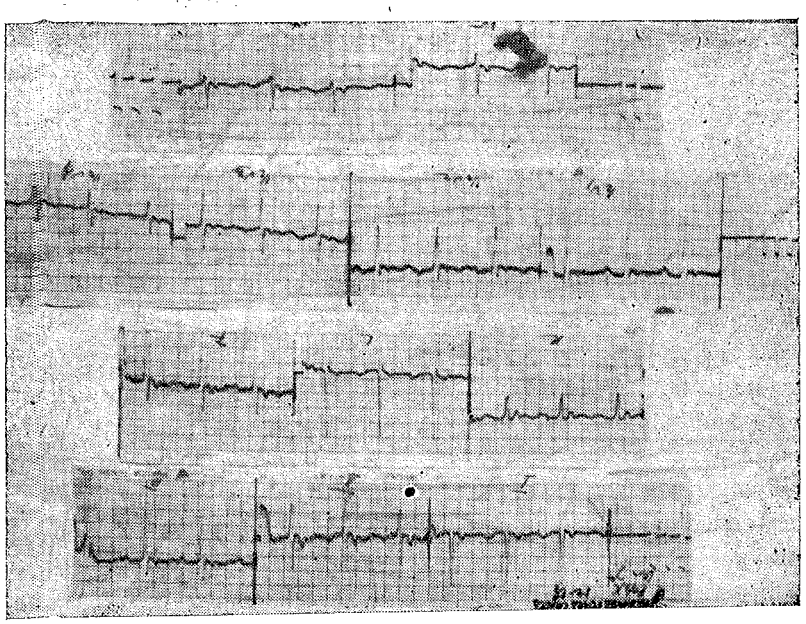
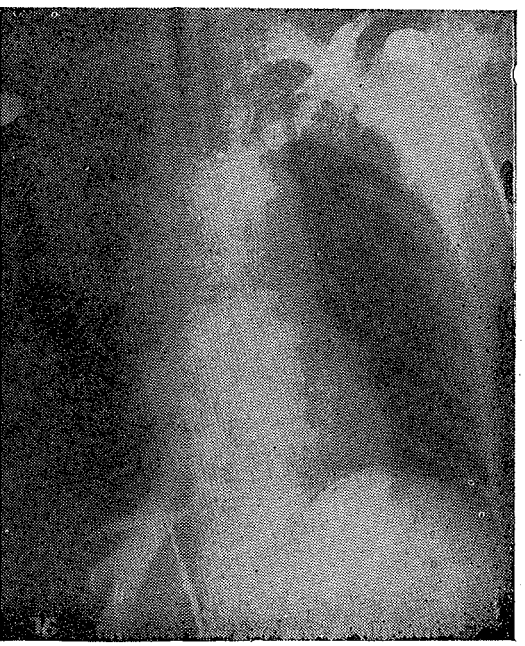


FIG 2: *ECG and 2 pieces of angiocardiographic pictures of case 2: Su. ♂, 6 months*





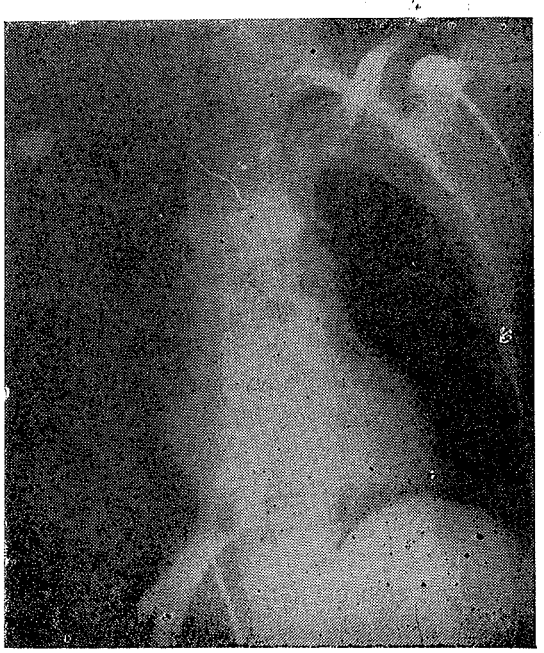


FIG 3 : *ECG and 2 pieces of angiocardiographic pictures of case 3 : J.K. ♂, 12 years*

On the basis of clinical findings as shown in table 1 the diagnosis of Tricuspid Atresia in the three described patients, Sa. ♀, 5, Su. ♂ 6/12, and J.K. ♂, 12, was highly suspected. All of them had even suffered from the complications of their primary disease neurologically, and ophthalmologically; two of them, Sa. and Su. had moderately to severely growth retardation mentally and physically, while the other, J K. had better growth pattern, mildly retarded, borderline IQ, and looked relatively well.

The hemodynamic studies performed to the three patients have comfirmed the diagnosis. The anatomical type of the lesion in pathient Sa. was very likely of type IIb, in patient Su. was type Ia, while in patient. J.K. was of type IIb, but with mild degree of Pulmonary/Subpulmonary Stenosis.

Two patients underwent surgical intervention; Sa, 5, type IIb, and Su. 6/12, type Ia. In patient Sa. the pulmonary artery was too small, its diameter was only 1/3 of that of the Superior Caval Vein, so that Glenn procedure could not be done — Waterston procedure was done instead. But unfortunately the patient died later of intractable heart failure and DIC (Disseminated Intravascular Coagulation) on the 10th day after surgery. On patient Su, Glenn procedure has been performed. But again unfortunately the patient died of cardiorespiratory arrest due to Vena Cava Superior syndrome on the same day of surgery.

## Discussion

The prevalence of Tricuspid Atresia in this study, found to be 0.93% of Congenital Heart Disease, is almost similar to the figures reported by various authors, around 1 - 3%, either from abroad (Dick et al., 1975; Diehl et al., 1968; Keith et al., 1967; Nadas et al., 1972; Kjellberg et al., 1955; Taussig et al., 1960) as well as from Indonesia (Hanafiah et al., 1965 : 2%; Maemunah et al., 1974 : 0.7%).

Although only 3 patients in this serie, underwent further hemodynamic studies, all the results showed that the clinical diagnosis could be considered as sufficiently reliable. But to determine the precise anatomical type of the lesion is indeed very difficult without selective angiocardiography and/or autopsy.
Even in the 3 catheterized patients, the important lateral view of selective angiocardiography could not be visualized because of some technical difficulties.
Moreover autopsy still cannot be done here due to difficulties to obtain permission. So, the types of anatomical lesion in this series were made on the basis of other data only, as proposed by various authors.

The clinical features of this disease depend much on the anatomical type of the lesion, and generally various complications are quite common, including cerebrovascular accidents that may lead to death, or residual hemiparesis, convulsive disorders and mental retardation. Phornphutkul et al. (1973) described that an aggresive surgical approach with early correction of the cardiac malformation in infants and children with cya-

notic Congenital Heart Disease may significantly reduce the incidence of this complication.

The prognosis of Tricuspid Atresia is said to be very poor. Without surgery prospects for long term survival are poor, and death is usual within the first year of life, especially in the group with diminished pulmonary blood flow. Shunt operations are reported to increase longevity, but to what extent is unknown. New surgical approaches such as right atrial-pulmonary arterial anastomosis and the Fontan procedure may result in a more prolonged survival.

The death of the 2 patients undergoing surgical intervention, were presumably, one due to complication of Glenn procedure, while the other due to Waterston procedure. This kind of surgical death was also encountered in every center abroad (Nadas et al.,1972; Tay et al., 1974). But the comparison of surgical/medical mortality between this clinic and other centers abroad could not be made because of the too small quantity of material here.

Anyhow we have to conclude that early recognition and appropriate surgical intervention should be stressed and still should be improved in connection with recent reports from abroad that the poor prognosis of this clinical entity, in that way, may be greatly improved.

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