ORIGINAL ARTICLE

Tetralogy of Fallot at the Department of Paediatrics, Medical Faculty, Hasanuddin University, Ujung Pandang

by

J.M.CH. PELUPESSY and HUSEIN ALBAR

(From the Department of Paediatrics, Medical Faculty, Hasanuddin University/ General Hospital, Ujung Pandang)

Abstract

A retrospective study was carried out on children with tetralogy of Fallot at the Department of Paediatrics, Medical Faculty, Hasanuddin University/Ujung Pandang General Hospital from January 1, 1982 to December 31, 1985 inclusive.

The diagnosis of tetralogy of Fallot was based on the history, clinical and laboratory findings, chest radiograms and electrocardiograms. There were 10 children included consisting of 7 in- and 3 out-patients, ranging in age from 17 days to 10 years. All in-patients had protein energy malnutrition and or respiratory tract infection. Boy to girl ratio was 1 : 1,5.

Cyanosis developed before the age of six months, clubbing of the fingers and toes manifested after one year of age while squatting became obvious after the age of two and a half years.

Introduction

The anatomical basis of cyanotic congenital heart disorder comprising pulmonary stenosis, ventricular septal defect, overriding of the aorta and right ventricular hypertrophy was first described about 200 years ago. These lesions which were defined for the first time by Fallot in 1888 as a syndrome, were then called "tetralogy of Fallot" (TF) (Jordan and Scott, 1973; Kawabori, 1979; Nadas, 1969).

This anomaly occurs at about 0,4 per 100 live births and becomes the most common form of cyanotic congenital heart disease after the first year of life (Jordan and Scott, 1973; Scott, 1987). According to several reports, the deformity accounts for at least 10% of all congenital cardiac anomalies (Kawabori, 1979). TF is more common in boys than girls (Scott, 1987).

The aim of this paper is to inform some clinical observations of patients with TF, especially on clinical features, laboratory findings, radiographic and electrocardiographic patterns.

Materials and Methods

The medical records of children with TF, hospitalized as well as outpatients at the Department of Paediatrics, Medical Faculty, Hasanuddin University/Ujung Pandang General Hospital from January 1, 1982 to December 31, 1985 inclusive were reviewed.

Criteria for the clinical diagnosis of TF were as follows :

(1) Clinical picture. Signs of cyanosis, heart murmur, clubbing of the fingers and toes and squatting. The child was defined cyanotic when it appeared blue at rest or when crying. Cyanosis was predominantly noted on the mucous membrane, nails and skin. Clubbing of the fingers and toes were marked by a drum stick appearance around the edge of the fingers and toes while squatting was a kneechest position made up as an attempt to reduce cyanosis and dyspnoea during physical exercise.

(2) Lobaratory findings revealed the presence of polyglobuli, noted when haemoglobin, erythrocyte and haematocrit values were above normal for age (Alter et al., 1981).

(3) X-ray films of the chest showed a relatively small heart of right ventricular contour and a poorly vascularized lung field (boot-shaped silhouette) (Nadas, 1969).

(4) Electrocardiogram demonstrated right axis deviation (RAD) and right ventricular hypertrophy (RVH).

Data of age and sex, nutritional status, evidence of cyanosis, heart murmur, clubbing of the fingers and toes, squatting, the presence of polyglobuli, chest radiograms and electrocardiograms were recorded. Nutritional status of the patients was determined based on the body weight to height parameter according to the Indonesian Standard (Staf Bidang Sosio-Ekonomik Gizi dan Statistik Direktorat Gizi Departemen Kesehatan R.I, 1978).

Patients with incomplete data were excluded from the study.

Results

Only 10 cases (50%) of 20 patients with cyanotic heart disease seen between January 1, 1982, and December 31, 1985, fulfilled the criteria for the diagnosis of TF. There were 7 (70%) in-patients and 3 (30%) out-patients, ranging in age from 17 days to 10 years.

Table 1 : *Age and sex distribution*

age group (years)	boys	girls	N	%
0 - 1	1	3	4	40.00
1 - 5	1	2	3	30.00
5 - 10	2	1	3	30.00
total	4	6	10	100.00

The majority of patients (70%) were underfives with a boy to girls ratio of 1 : 1,5.

Table 2 : *Distribution of nutritional status*

nutritional status	out-patients N	%	in-patients N	%	N	%
well-nourished	2	66.65	0	00,00	2	20.00
under-nourished	1	33.35	2	28,57	3	30.00
poorly-nourished	0	00.00	5	71,43	5	50.00
total	3	100.00	7	100,00	10	100.00

Table 2 shows that all in-patients suffered from PEM.

Table 3 : *Clinical and laboratory findings, chest radiograms and electrocardiograms*

Abnormalities	N	%
Cyanosis	10	100.00
Clubbing of fingers & toes	6	60.00
Squatting	4	40.00
Heart murmur	10	100.00
Respiratory tract infection	7	70.00
Polyglobuli	9	90.00
Boot-shaped silhoutte	10	100.00
ECG (RAD + RVH)	10	100.00

Table 4 : *Age distribution according to clinical onset of cyanosis*

Age of onset of cyanosis (months)	N	%
0 - 1	6	60.00
1 - 3	1	10.00
3 - 6	3	30.00
total	10	100.00

Table 5 : *Onset of clubbing & squatting according to age*

Age (yrs)	N	Clubbing of the fingers and toes N	%	Squatting position N	%
0 - 1	4	0	00.00	0	00.00
1 - 5	3	3	100.00	1	10.00
5 - 10	3	3	100.00	3	100.00
total	10	6	60,00	4	40.00

Cyanosis became obvious before the age of 6 months in all cases (see table 4). Clubbing of various degrees and squatting were not observed before the age of one year. Between 1-5 years, 3 patients demonstrated clubbing of the fingers and toes whereas 1 patient developed squatting at two and a half years of age. After the age of 5 years, all patients had already showed clubbing of the fingers and toes and squatting.

Discussion

The severity of pulmonary stenosis and ventricular septal defect determines the clinical manifestation of TF. The clinical picture varies from asymptomatic (pink tetralogy) or mild cyanosis to severe, leading to a neonatal emergency with profound cyanosis during the first week of life (Shinebourne and Anderson, 1980). Cyanosis as the main symptom, accompanied by clubbing and squatting and also failure to thrive, become more obvious as the child grows up (Jordan and Scott, 1973). Cyanosis is noted in about one third of infants at birth (Nadas, 1969).

Squatting usually becomes more evident when the child starts to walk varying from 1½ to 2½ years of age (Nadas, 1969). The presence of clubbing varies with the severity of the disease, usually after the age of 1 or 2 years (Jordan and Scott, 1974).

In our study, 60% of cases developed cyanosis in the neonatal period whereas the remainder became cyanotic before the age of 6 months (table 4). Clubbing of the fingers and toes manifested after one year of age, while squatting was noted after two and a half years (table 5).

Asymptomatic or mild cyanotic cases without any complications or other associated diseases are generally not hospitalized. All our in-patients were hospitalized due to protein energy malnutrition and/or respiratory tract infection.

Polyglobuli is an adaptive mechanism to lower arterial oxygen saturation because of a right-to-left shunt and is a very good indicator of hypoxia (Kawabori, 1978; Nadas, 1969).

TF is the most common type of cyanotic heart disease (Kawabori, 1978; Nadas, 1969). We found an incidence of 50%. Its prevalence was 7,35% of all observed congenital heart diseases (Endang and Pelupessy, 1986).

The ratio of boys to girls in our series was 1 : 1,5 (table 1) while Siregar et al. (1980), found 1, 17 : 1 and Teguh et al. (1981) 1, 2 : 1.

There were no deaths during the period of observation. Death rate is usually high in severe cases, mostly in the first year of life, which then decreases after the age of two (Jordan and Scott, 1973). The average age at death is 12 years (cited from Jordan and Scott, 1973). Teguh et al. (1981) in Surabaya noted a mortality rate of 25%.

Iron administration to patients with relative iron deficiency anemia may cause dramatic improvement of clinical symptoms (Jordan and Scott, 1973). Nadas recommended to keep the haematocrit level between 55 and 75%. If the value is lower than 55% (relative iron defiency anemia) then iron is indicated and when it is higher than 75%, partial exchange transfusion should be advocated (Black and Lubchenco, 1982).

Surgical intervention is indicated in patients with severe hypoxia, polyglobuli, hyperpnoeic spells and failure to thrive (Kawabori, 1978).

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