ORIGINAL ARTICLE

Factors Associated with the Occurrence of Cyanotic Spells in Tetralogy of Fallot Patients

Teddy Ontoseno

(Department of Child Health, Medical School,
University of Airlangga/Dr. Soetomo Hospital, Surabaya)

ABSTRACT A study was carried out on 114 tetralogy of Fallot patients attending the Department of Child Health, Medical School, University of Airlangga/Dr. Soetomo Hospital between 1 January 1988 to 31 December 1992. Only 81 patients fulfilled our study criteria where 52 (64.2%) were cases with complications such as cyanotic spells, 4 (4.93%) among them had brain abscesses. Twenty-nine individuals without complications acted as controls. Age, sex, nutritional status, hematocrit, MCHC and onset of symptoms between the two groups were analyzed using the multiple regression logistic. It has been shown that relative anemia, polycythemia and the age of 2-5 years contributed to the onset of cyanotic spells, respectively, R = 0.3171 and p = 0.0004; R = 0.2220 and p = 0.0073; R = 0.1363 and p = 0.00465. There fore, in conventional treatment of tetralogy of Fallot patients it is essential to observe these risk factors in order to avoid complications and to improve the quality of life in these patients who are on the waiting list for surgery.
[Paediatr Indones 1995; 35:227-230]

Introduction

Tetralogy of Fallot is the most commonly found type of cyanotic congenital heart disease.[1] Open cardiac surgery is the only definite treatment of patients with tetralogy of Fallot;[2] however, complex and sophisticated equipment and facilities are essential for this kind of expensive surgery, besides great experience and a good team-work among the specialists involved.[3,4] Thus, not all patients have the chance to be operated on; just few medical centers are able to perform this kind of surgery. Unfortunately, not all operated patients show the expected results.[5,6] On the other hand, although new cases are diagnosed every year, the majority of these patients come from a low socio-economic class, thus resulting in many

Accepted for publication: July 30, 1995. Author's address: Teddy Ontoseno, MD, Department of Child Health, Medical School, University of Airlangga, Jl. Darmohusodo, Surabaya, Indonesia.

unoperated tetralogy of Fallot patients. Furthermore, some patients on the waiting list for surgery who are being treated conventionally, often develop life threatening complications, which results in increased mortality rate and complications during surgery.[7] It is essential to find factors associated with the development of complications in patients who are being treated medically. If these factors can be controlled it would be possible to prevent these complications and a better quality of life can be expected for these patients while waiting for surgery.[8]

Methods

A case-control hospital-based study was carried out on tetralogy of Fallot patients from January 1988 to December 1992, consisting of outpatients and in-patients of the Division of Cardiology, Department of Child Health, Dr. Soetomo Hospital, Surabaya. Patients included in this study comprised all echocardiographically confirmed tetralogy of Fallot patients with complication, i.e., cyanotic spells which were evidenced a history of sudden deep and fast breaths which accompanied by increased cyanosis resulting in weakness, to fainting spells and convulsions, or those suffering from brain abscess indicated by fever, leukocytosis, increased intracranial pressure, or evidence of brain abscess on skull x-ray and CT-Scan. Tetralogy of Fallot patients without complications served as controls.

Laboratory criteria were as follows: polycythemia was defined as hematocrit $\geq$ 65% with hemoglobin level $\geq$18 g/dl, relative anemia was defined as MCHC < 30%. The age was based on the birth certificate at the time of admittance, the nutritional status was grouped on age matched body weight according to the NCHS/WHO. The nutritional status was considered as good if it was within 2 SD from the standard, medium if 3 SD from the standard, poor if 4 SD from the standard. The time at which the patient for the first time complained about complications was considered as the time of the first complaint.

Multiple logistic regression method was used to predict the probability of complications as a consequence of several risk factors and determine the odds ratio.

Results

The subjects consisted of 81 (71.1%) patients fulfilling the study criteria, consisting of 47 boys and 34 girls and 29 (35.0%) controls. Of the 81 patients, 52 (64.2%) experienced cyanotic spells and 4 (4.9%) had a brain abscess besides cyanotic spells. The age of the patients ranged from 3 months to 12 years with 19.8% under 2 years old, 39.5% between 2-5 years, and 40.8% over 5 years old.

The cyanotic spells mostly occurred between 2-5 years (46.2%); the prevalence of the spells was higher in boys than in girls (1.26:1).

In most cases, the hemoglobin concentration was more than 18 g/dl and 42.3% of the patients showed a hematocrit of more than 65%. Brain abscess was found at the age of more than 3 years (75%) with boy to girl ratio of 3:1, and hematocrit of more than 65%.

A good nutritional status was found in 24.7% of the cases and only 17.3% of the cases were accompanied by cyanotic spells 25% of the cases were accompanied by brain abscess. A poor nutritional status was found in 75.3% of the cases and 82.7% suffered from cyanotic spells, 75% cases were accompanied by brain abscess.

Relative anemia was found in 51.8%, of which 73.1% suffered from cyanotic spells. All brain abscess cases were accompanied by relative anemia. Polycythemia was found in 29.6% of the cases.

Hemoglobin concentration, hematocrit, relative anemia and age 2-5 years showed a significant association with cyanotic spells. As seen from the logistic regression analysis the probability of tetralogy of Fallot patients to develop cyanotic spells is:

$$P(X) = \frac{1}{1+e(-3{,}0121+2{,}89 \times \text{relative anemia} + 2{,}8678 \times \text{polycythemia} \times \text{age(2-5 year)}}$$

A rating of 1 point is given if the above mentioned symptoms occur and zero point if no symptoms occur. P(x) is the probability of a tetralogy of Fallot patient to develop cyanotic spell, as the exponential value or natural logarithmic value = 2.718.

Discussion

Relative anemia (p=0.0004 and r = 0.3170) which is found in 51.9% of the patients contributed the most to cyanotic spells. The odds ratio of relative anemia was found to be 18.0769; thus, the risk of tetralogy of Fallot patients with relative anemia experiencing cyanotic spells is 18 times more than patients without relative anemia.

Following relative anemia, the hematocrit levels played a great role in the occurrence of cyanotic spells. The hematocrit levels showed a significant positive correlation with cyanotic spells (p=0.0073 and r = 0.222) and showed that the relative risk in the occurrence of cyanotic spells was 17.6 times greater than without polycythemia. The higher the hematocrit levels, the greater the blood viscosity and consequently resulting in an increase in disturbances of the blood flow, especially in the circulation to the brain.

It has been observed that age of the patient has a significant correlation with cyanotic spells, 2-5 years of age (p= 0.0465, r = 0.1363 and OR = 6.7864). At the age of 2-5 years the relative risk of developing cyanotic spells was 6.8 times greater than the other age groups. According to child, 1991, the hemodynamic changes are evidence at 2-4 year in the form of hypoxia of various degrees.[7] According to Sastroasmoro[9] cyanotic spells decrease in frequency as well as intensity following the formation of blood vessel collateral at the age of 18 moths to 2 years.

Previous investigators observed that the prevalence of brain abscess was 23%[7] however, in this study it was shown to be only 4.9%. This appears to be caused by the fact that many patients with a brain abscess did not visit the hospital and that this investigation was only limited to patients aged 12 years old while the occurrence of brain abscess is mostly at the age of more than 12 years.[7] No significant

correlation was found in patients with brain abscess with other parameters, this may be caused to the relative small number of patients studied. However, it has been observed that the mean hemoglobin in patients with brain abscess was 19.4500 ± 1.23234 and the mean hematocrit was 68.75 ± 4.8563.

Sastroasmoro[9] noted that the prevalence of brain abscess or CVA was higher in patients with hemoglobin and hematocrit levels of more than 18 gr/dl and 65%. This is accordance with the results of this study: mean hemoglobin in the group without complications of 17.9207 ± 1.7498, and 19.4500 ± 1.2234 in the group with a brain abscess, and a mean hematocrit of 68.75 ± 4.8563.

Conclusions

From the results of the study it has been shown that relative anemia, polycythemia, and age 2-5 years are associated with the occurrence of cyanotic spells. It is possible that other factors may also be involved, however, other parameters such as platelet count, albumin concentration and nutritional status have not been studied. By using the multiple logistic analysis the probability of Tetralogy Fallot patients in developing cyanotic spells can be calculated as follows:

$$P(X)=\frac{1}{1+e(-3{,}0121+2{,}89 \times \text{relative anemia} + 2{,}8678 \times \text{polycythemia X age(2-5 year)})}$$

Further investigations have to be performed in order to study other risk factors which may be associated with cyanotic spells, brain abscess or other complications.

By taking the risk factors into consideration it is possible to avoid complications during the pre-operative preparations in order to obtain optimal results.

References

1. Jordan SC, Scoot O. Tetralogy of Fallot. In: Heart disease in pediatrics; 3rd ed. London: Butterworths 1989; 148-57.
2. Graham TP. When to operate on the child with congenital heart disease. Pediatr Clin North Am 1984; 31:1275.
3. Tahalele P. Permasalahan operasi jantung di RSUD Dr. Soetomo Surabaya. Warta IKABI, 1988; I: 49-53.
4. Stephenson LW, Friedman MD, Edmunds LH. Staged surgical management of tetralogy Fallot in infants. Circulation 1978; 58: 837.
5. Prasodo AM. Cyanotic congenital heart disease without surgical intervention. Maj Kardiol Indones 1988; 3:119-23.
6. Anderson RH Macartney FJ, Shinebourne EA, Tyan M. Pediatric cardiology, Vol. 2. Edinburg: Churchill Livingstone, 1990;765-93.
7. Ontoseno T. Serangan sianosis. Continuing Education Ilmu Kesehatan Anak, 1991; 23: 91-101.
8. Jordan SC, Scott Olive. Complications of Congenital Heart Disease. Heart Diseases in Paediatrics. Third ed. Butterworths London, Boston, Singapore, Sydney, Toronto, Wellington, 1989; 262-73.
9. Sastroasmoro S, Madiyono B. Penyakit jantung bawaan sianotik dengan vaskularisasi paru berkurang. In: Markum AH et al. eds. Buku ajar ilmu kesehatan anak, vol I. FKUI Jakarta, 1991, 577.