Cooperation between Pediatric Cardiologists and Cardiac Surgeons for Congenital Heart Diseases

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Cooperation on a 24-hour basis between pediatricians, surgeons, nurses and paramedical staff is extremely important for the management of severe congenital heart diseases. It is also of the utmost importance for the cardiac surgeons to provide a precise and correct diagnosis of cardiac morphology and function by non-invasive or invasive methods in patients before surgery.

The surgical results have been improved every year, but the post-operative prognoses of patients with complete transposition of the great arteries with pulmonary hypertension, Taussing-Bing's anomaly, double outlet right ventricle and neonate congenital heart diseases are still grave. Progress in surgical techniques and elimination of errors in surgical procedures and treatments of postoperative complications will improve surgical results.

The prevention and management of cardiac failure or arrhythmia after surgery are important for patients with these problems. The improvement of surgical techniques is desirable especially to prevent residual disorders and complications after surgery.

Recently there has been a decrease in natural cardiac deaths, but there has been no particular change in the number of deaths from myocardial disease or complex cardiac lesions. New approaches for treating these patients are a matter for future consideration.

The number of patients with congenital heart disease has been decreasing markedly every year due to the decrease in the birth rate in Japan, but doctors still must treat the lives of diseased children even more carefully than they do at present and must continue their efforts to help these patients lead normal lives by the complete correction of their diseases.

A close cooperation between pediatricians and surgeons and the teamwork of doctors, nurses and paramedical staff are extremely important in the treatment of severe congenital heart disease (CHD).

The prognosis of neonates with CHD becomes progressively worse as time goes by. In the previous study, among all the neonates with CHD who died, death occurred within 2 weeks after admission except in one case: one half of these (27/51 = 52.9%) died within 3 days and 41.1% (21 cases) died following surgery within 9 days after admission.1

Neonates with CHD must be treated promptly by a pediatric cardiologists and cardiac surgeons. However, urgent cardiac examinations and treatments cannot succeed without the cooperation of the hospital staff. Although surgical procedures at midnight may be postpone with prostaglandine E1 therapy, surgical treatment cannot be delayed for long.

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SUBJECTS AND METHODS

The subjects of the present study were 2,062 children, including 245 neonates, who were admitted to National Children's Hospital, Tokyo, for congestive heart failure or hypoxemia, requiring surgical intervention during a period from January, 1976 to December, 1981.

The mortality or the survival rate of these patients was calculated according to their age and diagnosis.

RESULTS

There occurred 316 deaths including 115 neonates (36.4%) of 0–28 days old. Among these 316 cases 233 (73.7%) were under one year of age (including the neonates).

Surgical intervention was carried out for 856 cases including 62 neonates. There were 189 deaths after surgery including 41 neonates (Fig. 1).

The overall mortality rate was 15.3% (316/2062). Neonates accounted for about 1/3 of all deaths (115 cases) and cases under one year of age (including the neonates) about 3/4 (233 cases). The mortality rate of the neonates was especially high (115/245 = 46.9%). The incidence of surgical death was 22.1% (189/856) as a whole and 66.1% (41/62) in the neonates. The incidence of non-surgical death was 10.5% (127/1206) as a whole and 5.2% (53/1023) in cases excluding the neonates.

Figure 2 shows the mortality age distribution.
The incidence of death in patients with CHD decreased with age, and it was especially low over 2 years of age.

Figure 3 shows the diagnosis and age distribution of the fatal cases.

The prognosis of coarctation complex, total anomalous pulmonary venous connection, pure pulmonic atresia and so on were especially poor irrespective of surgical intervention. The incidence of non-surgical death decreased after the neonatal period except in cases with myocardial diseases and with complex cardiac defects, such as asplenia syndrome, polysplenia syndrome, hypoplastic left heart syndrome and so on.

The outcome of surgical procedures has been improved every year, but is still poor for cases under one year of age with complete transposition of the great arteries, Taussig-Bing's anomaly and double outlet right ventricle, and for neonatal CHD. Some infants with ventricular septal defects were lost due to complications.

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**Fig. 3. Diagnosis and age distribution of fatal cases.**

- **PA** = pulmonic atresia; **PS** = pulmonic stenosis; **TAPVC** = total anomalous pulmonary venous connection; **TGA** = complete transposition of great arteries; **DORV** = double outlet right ventricle; **CAVO** = common atioventricular orifice; **VSD** = ventricular septal defect; **ASD** = atrial septal defect; **PDA** = patent ductus arteriosus; **HLHS** = hypoplastic left heart syndrome

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**Fig. 4. Number of births in Japan as a whole and in Tokyo (upper panel) and changes in the number of cardiac patients at outpatient clinics (lower panel) during a period from 1970 to 1980.**
In cases over one year of age, poor surgical results were found in tetralogy of Fallot (including extreme type), common atrioventricular orifice, double outlet right ventricle, single ventricle, complete transposition of the great arteries, ventricular septal defect complex and so on.

DISCUSSION

The main causes of surgical death are under-estimation of obstructive changes of the pulmonary arteries, delay of surgical procedure for tricuspid, mitral or aortic stenosis, persistence of pulmonary hypertension due to collateral circulation and existence of multiple VSD.

It is important to estimate ventricular volume, and cardiac morphology and function, especially in cases associated with valvular heart disease.

The prognosis of CHD will be improved markedly if errors in surgical techniques and mismanagement after surgery can be eliminated.

Present surgical interventions can save many cases with CHD. Future developments in surgical procedures will increase the number of curable cases.

However, it is difficult to decide the appropriate treatment for cases with complex cardiac lesions, i.e., medical or surgical treatment.

Heart failure and arrhythmia are critical conditions which occur frequently after surgery. Heart failure due to pulmonary stenosis and/or pulmonary regurgitation following intracardiac correction of tetralogy of Fallot or due to residual shunt are difficult to control.

Arrhythmia following intraatrial manipulation, such as in Mustard’s or Senning’s operation, can be fatal. Thus, the prevention and management of arrhythmia is considered most important.

It is also important to improve surgical procedure so as to correct cardiac anomalies as completely as possible in order to give patients a chance to lead a normal life. Although the birth rate in Japan has been decreasing markedly every year (Fig. 4) as well as the number of patients with CHD, doctors still must treat diseased children even more carefully than they do at present.

REFERENCE