Tetralogy of Fallot

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According to WHO estimates, 16.7 million people around the globe die of cardiovascular diseases each year.[1] **Among** the total CVD deaths annually, about 8.6 million are of women. Heart attack and stroke deaths are responsible for twice as many deaths in women as all cancers combined.[2] In developing countries twice as many deaths from CVD now occur.[3] A particular cause for concern is the relatively early age of CVD deaths compared with those in the developed regions. By 2010 CVD is estimated to be the leading cause of death in developing countries.[4] By 2020 the WHO estimates nearly 25 million CVD deaths worldwide.[6] Heart disease has no geographic, gender or socioeconomic boundaries.

Between 1990 and 2020, deaths from non-communicable diseases and injury are expected to rise from 33 million to 58 million annually, with a similar proportional increase in years of life lost.[6] By 2020, cardiovascular diseases, injury and mental illnesses will be responsible for about one half of all deaths and one half of all healthy life years lost, worldwide.[7]

In Nepal, around 15 percent of the total population, numbering about 4 million, is suffering from heart disease, a press release issued by the Ministry of Health and Population (MoHP) and further said "On one hand, there is an increasing unhealthy lifestyle, eating habits, unbalanced diets. On the other, physical exercise is lacking, leading inevitably to heart disease," the release noted. MoHP has been coordinating with the Nepal Heart Disease Relief Academy and the members of Nepal Heart Network to provide treatment and preventive measures to fight against heart disease. World Heart day is observed every year on September 25. This year 2012, the slogan for World Heart Day is "Healthy Weight for Healthy Heart". MoHP also organized varied programs to create awareness about the disease on the World Heart day. MoHP has appealed to all citizens to eat a balanced diet, and exercise regularly to control weight as excessive weight is the major cause of heart disease. [4]

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Nepali people suffer from three types of heart diseases; Congenital, Rheumatic and Coronary heart diseases. The incidence of CHD was 5.8 per thousand hospitalized children.[9] The prevalence of rheumatic heart disease and congenital heart disease among school children of Kathmandu is 1.2/1000 and 1.3/1000, respectively, with mitral regurgitation and atrial septal defect being the commonest lesions. [10]

TOF is the most common cyanotic congenital heart defect and is estimated to account for 4% to 9% of congenital heart defects overall, or in the range of 0.262 to 0.392 per 1000 live births.[2] Tetralogy of Fallot was first described by Niels Stenson in 1671, although its precise anatomical description was elegantly illustrated by William Hunter at St Georges Hospital Medical School in London in 1784. His description of a large outlet ventricular septal defect together with subpulmonary and pulmonary valve stenosis, and its resulting physiology, was refined by Etienne-Louis Fallot in 1888 in his description of *L'anatomie pathologique de la maladie bleu*, but the term tetralogy of Fallot (a tetrad of (i) ventricular septal defect with (ii) over-riding of the aorta, (iii) right ventricular outflow obstruction, and (iv) right ventricular hypertrophy) is attributed to Canadian Maude Abbott in 1924.

Study from Dhukhekl hospital found, Tetralogy of Fallot (TOF) in 13.1%,of cyanotic heart disease, similarly total anomalous pulmonary venous connection (TAPVC) 3.6%, transposition of great arteries (TGA) with VSD 1.2% and unspecified cases of heart disease in 13.1%. VSD and TOF were the most common lesions while other CHD like ASD, dextrocardia, TAPVC, ECD, TGA with VSD were encountered less frequently. The mortality rate was 20 %. [9]

The 4 features typical of Tetralogy of Fallot include right ventricular (RV) outflow tract obstruction (RVOTO) (infundibular stenosis), ventricular septal defect (VSD), aorta dextroposition, and right ventricular hypertrophy. Occasionally, a few children also have an atrial septal defect (ASD), which makes up the pentad of Fallot. The basic pathology of tetralogy is due to the underdevelopment of the right ventricular infundibulum, which results in an anterior-leftward malalignment of the infundibular septum. This malalignment determines the degree of RVOTO.

The cause of most congenital heart defects is unknown. Many factors seem to be involved. However Factors that increase the risk for this condition during pregnancy include: Alcoholism in the mother, diabetes, mother who is over 40 years old, poor nutrition during pregnancy, rubella or other viral illnesses during pregnancy

Children with tetralogy of Fallot are more likely to have chromosome disorders, such as Down syndrome and DiGeorge syndrome (a condition that causes heart defects, low calcium levels, and poor immune function).

Signs and tests

A physical examination with a stethoscope almost always reveals a heart murmur. The relevant test may include: Chest x-ray, Complete blood count, Echocardiogram, Electrocardiogram, MRI of the heart (generally after surgery)

Nonetheless, most children present with the condition after birth. Although an experienced paediatrician or cardiologist usually suspects the diagnosis clinically, transthoracic cross-sectional echocardiography provides a comprehensive description of the intracardiac anatomy. With the exception of patients with major aortopulmonary collateral arteries and rare cases in whom echocardiographic assessment is incomplete, any other diagnostic investigations (eg, cardiac catheterisation) are now rarely done before palliative or corrective surgery.

Treatment

Surgery to repair tetralogy of Fallot is done when the infant is very young. Sometimes more than one surgery is needed. When more than one surgery is used, the first surgery is done to help increase blood flow to the lungs.

Surgery to correct the problem may be done at a later time. Often only one corrective surgery is performed in the first few months of life. Corrective surgery is done to widen part of the narrowed pulmonary tract and close the ventricular septal defect.

Expectations (prognosis)

Most cases can be corrected with surgery. Babies who have surgery usually do well. More than 90% survive to adulthood and live active, healthy, and productive lives. Without surgery, death usually occurs by the time the person reaches age 20.

Patients who have continued, severe leakiness of the pulmonary valve may need to have the valve replaced.

Regular follow-up with a cardiologist to monitor for life-threatening irregular heart rhythms (arrhythmias) is recommended.

Complications

Few important complications are: Delayed growth and development, irregular heart rhythms (arrhythmias), seizures during periods when there is not enough oxygen and Death

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