

# **ABNORMAL HEART FROM BIRTH**

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# Abnormal heart from birth: What parents should know

By Dr. Julius A. Ogeng'o

## INTRODUCTION

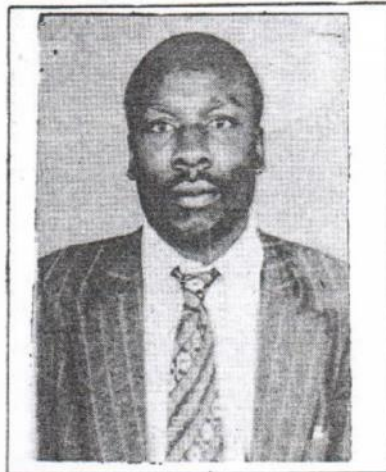
**E**ight to thirteen in every 1000 of all babies born have one form or another of developmental defects of the heart, hereafter referred to as congenital heart disease. While some defects may be minor and course no symptoms, others may be severe enough to kill soon after birth. Between these two extremes, are a whole array of defects that will manifest at various times between birth and adulthood, in different ways. These defects can occur singly or multiply in various combinations.

Indeed studies done in Kenyatta National Hospital, between 1975 and 1980 showed that over 53% of the children seen in the child Heart Clinic had congenital heart disease and that more than 60% presented in the first two years of life. However, due to the tremendous advances in the diagnosis and management of congenital heart disease, many patients with complicated congenital heart disease are now surviving up to adulthood.

Before discussing the details of congenital heart disease, the basic events in the development of the heart should be understood and readers interested in more than is provided may consult the editor.

## MAJOR EVENTS IN THE DEVELOPMENT OF THE HEART

The heart develops during the



Dr. Ogeng'o: "Eight to thirteen in every 1000 of all the babies born have one form or another of developmental defects of the hearts."

third week and starts pumping as early as the 21st day after conception which is well ahead of the other organs. The critical period is between the 3rd and 9th weeks and the processes are numerous and rapid.

It develops as a primitive tube called the cardiac tube that has two ends the venous end to which blood returns and the arterial end from which blood leaves.

When developing baby folds, the heart tube also folds so that the venous end lies behind the arterial end.

The primitive tube is then divided into a left and right sides. Subsequently each of these is divided into two. This series of divisions results in four chambers, on the venous end called cleft and right atrium, and on the arterial end called right and left ventricle.

Initially, a single trunk leaves the ventricles but later it is partitioned into two; the pulmonary trunk, leaving the right ventricle to supply the lungs and the aorta leaving the left ventricle. The origin of these two main arteries is guarded by valves which ensure that blood does not flow back to the heart. Valves also develop at the openings between each atrium and the corresponding ventricles and ensure that blood only flows from the atrium to the ventricle and not the reverse.

In the unborn child, the lungs are not functional and blood from the pulmonary trunk to the aorta through a conduct called *DUCTUS ARTERIOSUS*

## IN THE UNBORN BABY

The placenta is the organ which provides for the exchange of oxygen and carbon dioxide. The baby receives oxygenated blood from the mother through the placenta and via the umbilical veins. This oxygenated blood together with unoxygenated blood from the trunk and legs gets to a vein called the *INFERIOR VENA CAVA* which carries most of the blood to the right atrium. From here, the blood goes to the left atrium through an oval opening between these two atria (atria-plural for atrium). The remainder of the blood, together with blood from the head, neck and arms goes to the right ventricle and from here to the pulmonary artery. This blood is shunted to the aorta via the *DUCTUS ARTERIOSUS* and the little that may go to the lungs returns to the left atrium via the

pulmonary veins.

Blood from the left atrium goes to the left ventricle and from here through the aorta to the umbilical veins and from there to the placenta. Some of this blood goes to the head, neck and arms.

### AFTER BIRTH

Placental circulation stops and the following changes occur:

- The lungs become functional and blood from the right ventricle goes there for oxygenation and removal of carbon dioxide.
- the oval opening between the two atria closes.
- the *DUCTUS ARTERIOSUS* closes.
- the umbilical arteries and veins close.

### THE DEVELOPMENT DEFECTS

Any deviation from the normal pattern of the numerous, elaborate and rapid development processes may lead to defects of various types which can broadly be said to be due to one or more of the following.

- Failure of Some of the changes discussed above to occur after birth.

This causes persistence of circulation as it was in the unborn baby in spite of the absence of the placenta;

- Imperfect partitioning of the chambers leading to abnormal "holes in the heart";
- Narrowing or complete blockage of some of the circulatory channels;
- Imperfect development and or total absence of some of the parts.

### ASSOCIATED CAUSATIVE FACTORS

Although the exact cause of congenital heart disease is not known, the majority of cases are thought to be due to a complex interaction between environmental and hereditary factors. There is a greater risk of affected first degree relative. If one parent has a congenital heart disease, the chance

of the offspring having the same defect is approximately two to 4%. Inter marriages between close relatives may thus multiply the risk. When there is an underlying genetic disorder, the baby commonly has defects in other parts of the body. Therefore, since increasing maternal age is thought to increase the chance of genetic disorders, maternal age over 35 years may be associated with increased chances of congenital heart disease.

Environmental factors highly suspected to cause congenital heart disease include:

- Infections: While any infection during the 1st three months of pregnancy may carry a risk, it is estimated that about 10% of congenital heart defects may be due to German measles.
- Drugs: The list of drugs known to cause congenital defects in different parts of the body is long. Such drugs are referred to as teratogenic drugs and are generally not prescribed by doctors during the first three months of pregnancy. Some herbal medicines may fall in this category.
- Radiation: Accidental exposures to radioactive, paints, labels, fall outs or in the form of x-rays used for treatment and diagnosis of disease is dangerous in the first three months of pregnancy.
- Nutrition: Though no particular nutritional deficiency is known to cause congenital heart disease, malnutrition during early pregnancy may be an important factor in causation.
- Other diseases like diabetes melitus may be involved.

### DIAGNOSIS BEFORE BIRTH

With the advent of a specialised type of ultrasound called 2-D real time ultrasound; increasing attention has been paid to the diagnosis of congenital heart diseases and disorders of heart rhythm in the unborn child. By this means, most of the defects likely to cause problems can be detected between the 4th and 6th months after conception.

Unfortunately such means are expensive and very much limited to large specialised centres. However, when and where possible, the following should raise suspicion and the need to test:

- parents of a previously affected child
- severe growth retardation of the unborn baby.
- maternal age over 40 years
- maternal diabetes melitus
- exposure to German measles virus during early pregnancy
- excess fluid in the womb
- excess fluid in the abdominal cavity of the unborn baby

### SIGNS AND SYMPTOMS

Congenital heart diseases present as simple or as multiple and complex defects.

The most common modes of presentation are:

- Cyanosis; This refers to blue colouration best seen on the lips, tongue, eyes, fingers and toe nails. The defects that cause significant cyanosis are called cyanotic heart diseases and some of them may present with "hypercyanotic spells". In this condition, a child is normally mildly cyanosed, while playing becomes breathless, deeply cyanosed and prefers to squat. This may subside but is sure to recur. Victims of this condition may also present with finger and toe clubbing, gross enough to give them a drumstick appearance; recurrent chest infections; breathlessness on even mild exertion; poor exercise tolerance, headaches, a feeling of wanting to vomit or actual vomiting, blurred vision and brain function deficiency.
- Heart failure; This means that the heart is not able to sufficiently meet the body demands for blood. In older children, this is similar to adults but in babies the features may closely resemble those of other chest diseases. They include breathlessness and very rapid breathing even when on

breastfeeding.  
**Rapid or slow heart rate; swelling of the legs and liver.**

- Disturbances of heart rate: In the newborn, many stimuli can increase the heart rate to 200 beats per minute but rates exceeding this usually suggest a heart abnormality. There may also be audible heart beat or slowing down of the heart rate.
- Murmurs: These are abnormal sounds in the heart which only the doctor can hear with his special instrument called the stethoscope. While some of them do not signify disease, the doctor usually tells which ones are likely to signify congenital heart disease.
- Miscellaneous: These include poor feeding, retarded mental and physical growth with delayed milestones, abnormal sweating and general dullness.

## DIAGNOSIS

Having taken a clear history from the mother about the complaints, events during pregnancy, feeding habits, milestones, immunization and other diseases in the family, the doctor evaluates the general status of the child's health and examines the heart and blood vessels. Using the stethoscope, the doctor listens to the heart keenly and in 80% of the cases, he should make diagnosis.

He may then resort to investigations to confirm the exact disorders and assess the extent of the problem. Such investigations include chest x-rays and electrical activity of the heart which are routinely done and other specialised investigations like Echocardiography and cardiac catheterization which are only possible in the big hospitals.

## TREATMENT

Congenital defects which are asymptomatic and therefore discovered incidentally during the doctor's routine examination do not need treatment. Instead, they deserve close follow up. On the other hand, some of the serious

defects are amenable to successful surgical correction and this is the only definitive form of treatment. When such are diagnosed early, they can be surgically corrected and the affected child given a full quality and span of life.

## COMPLICATIONS

Congenital heart disease is capable of causing irreversible damage to the structure of the heart even in the parts which are initially normal. Besides, some of the defects predispose the heart to infections which may soon or later cause damage to parts of the heart, making it grossly inefficient. The primary congenital defect, the secondary damage to the heart and the long term effects of infections are known causes of heart failure.

Because of abnormal blood flow in many of these defects, the lungs may undergo progressive damage to

an eventual irreversible stage. The mixing of oxygenated and deoxygenated blood due to the defects in the heart and the secondary lung damage impairs the oxygen supply to the body. Thus, a situation in which the impaired heart function coexists with inadequate oxygenation of the blood contributes additively to jeopardise body function. This may in part explain why children with heart disease show mental and physical retardation. Some may even show obvious structural deformities.

In an attempt to compensate for the reduced oxygen levels, the red blood cells increase in number and in their level of the oxygen-carrying pigment. This renders the blood very viscous, reducing the rate of flow and this may cause problems like *STROKE* (discussed in the March/April 1989 issue).

For those who live to child-bearing age, there is increased risk of spontaneous abortion, pre-

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