



GUIDELINES FOR PARENTS

Care of a Child with Congenital Heart Disease



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10 FAQs on CARE OF A CHILD WITH CONGENITAL HEART DISEASE

1. My neighbor's son is suffering from heart disease who is only 6 months old. As a mother how will I know that my baby is also having heart disease or not?
2. My daughter is 2 years old. When I took her to a doctor, he suspected congenital heart disease and advised echocardiography.
 - (a) What is congenital heart disease?
 - (b) Will echocardiography confirm the doubt? Will it be harmful for her?
3. Can a baby with congenital heart disease lead a normal life? What are the precautions I should take for my baby?
4. My neighbor's son has been diagnosed to have a serious heart disease at the age of 3 months though he was absolutely normal at birth. Why were they not told that he has a heart disease at birth?
5. My pediatrician says that my baby is a blue baby. I only appreciate blueness of lips, palm, and soles while he cries. What exactly a blue baby is? How should I care for my baby? Is there a future for these blue babies?
6. My son is very active but during school routine screening suspected that he might be having some form of congenital heart disease which came out to be narrowing of a valve and need restriction of outdoor sports. I am very worried. How will I be able to manage my child? Doctor also talked about ballooning of valve, how does it help?
7. My baby is only 2 weeks old, and has been diagnosed to have congenital heart disease which will need surgery. I am very scared of surgery. Is there any medical treatment also available? What will be the quality of his life if surgery is done?
8. My 1-year-old baby has been diagnosed to have a hole in heart for which a device closure or surgery has been recommended. What should I do? If I go for a device how safe is it?
9. My first child, now 2 months old, is diagnosed to have some complex heart disease and will need multiple surgeries as advised by doctor. Is there any alternative of these surgeries for him? What all activities he will be able to do if all surgeries are successful? When should I plan for a second baby, is there a chance that my second baby will also have CHD?
10. My 10 days old baby suddenly became sick and the doctor said that he had coarctation of aorta. I do not want surgery for my small baby. My friend's son had aortic stenosis and underwent ballooning at the age of 4 days. He is doing well. I also want ballooning only. What must I do?

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Care of a Child with Congenital Heart Disease

PARENTAL GUIDELINES: FREQUENTLY ASKED QUESTIONS

INTRODUCTION

- The heart pumps blood to all the different parts of the body, supplying them with oxygen and energy. The heart sits between the lungs and works closely with them to make sure there is enough oxygen in the blood.
- A normal heart has the two atria (right atrium and left atrium) and two ventricles (right ventricle and left ventricle). There is a wall in-between two atria (interatrial septum) and two ventricles (interventricular septum) (**Fig. 1**).
- There are two lungs: left and right. In the lungs, carbon dioxide travels from the blood to the air and oxygen travels from the air to the blood.
- The oxygen-depleted blood from body returns to right atrium of heart through superior and inferior vena cava and then enters into right ventricle. Right ventricle receives, and pumps it down to the pulmonary artery and lungs to expel the carbon dioxide. The right ventricle has thinner wall than left ventricle.
- Atmospheric oxygen enters into lungs and then diffuses through alveolar membrane into the capillaries.
- Right and left pulmonary veins collect this oxygenated blood from respective lung lobes and bring it back to left atrium.
- This blood enters into left ventricle and then pumped down into aorta and get distributed to the various part of body.
- Usual systemic blood pressure is about 100–120 mm Hg (systolic) and 60–89 mm Hg (diastolic). Lung pressures are only one-fifth of systemic pressure.
- Average SpO_2 in right heart is 60–65% and on the left side SpO_2 is >95%.
- Normally, blood has only one pathway to enter into the left heart, that is, through the pulmonary circulation. Inside the mother's womb, lungs are like collapsed balloons within the amniotic fluid hence alternative channels—patent foramen ovale and patent ductus arteriosus are found. These channels must close after the birth. If they remain open, then an intervention may be required. In children with complex heart diseases, they become lifeline. Sometimes intervention is done to keep them open.

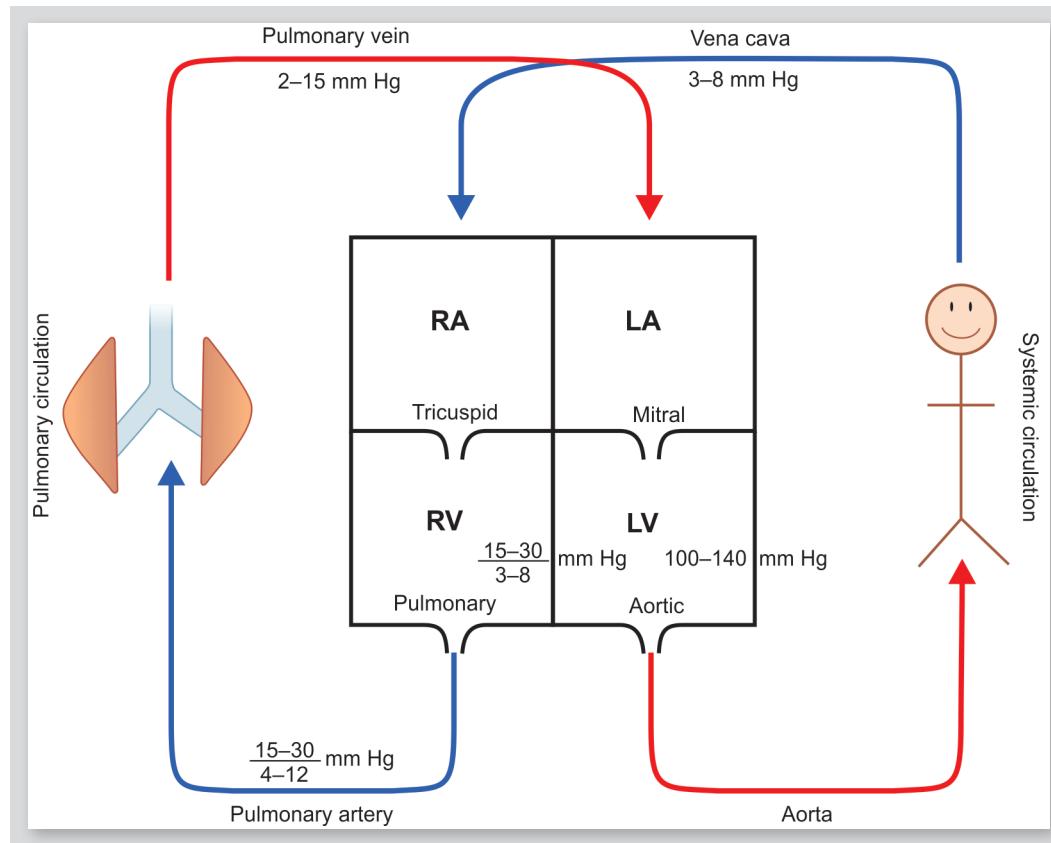


Fig. 1: Circulation of Heart: Pulmonary and systemic circulation.
(LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle)

The Square above represents the Heart

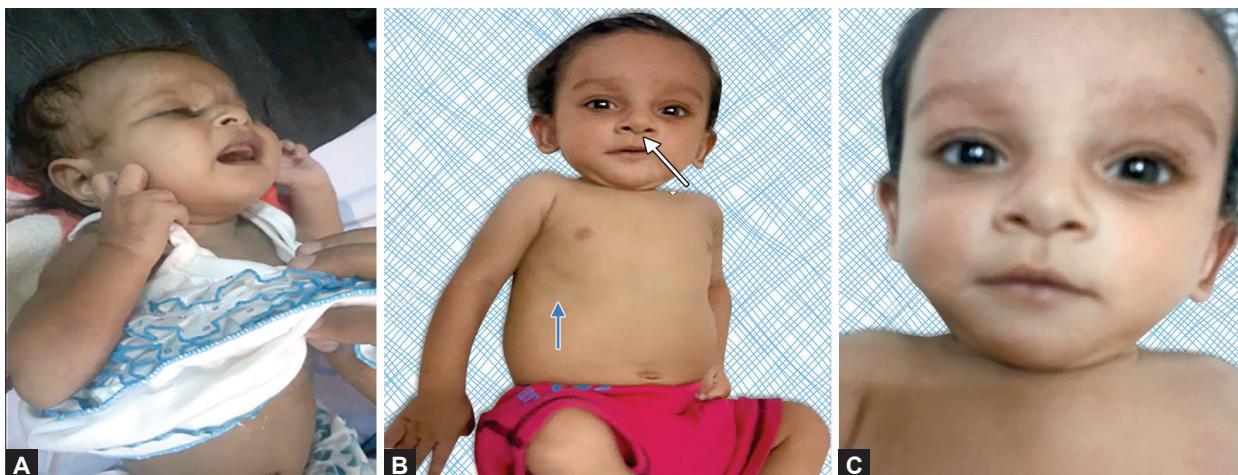
Q1

My neighbor's son is suffering from heart disease who is only 6 months old. As a mother how will I know that my baby is also having heart disease or not?

Heart disease can be detected in a child at any age. There are some symptoms which if present strongly favor heart disease. Symptoms vary according to age also but following are the red-flag signs which if present you must consult pediatrician and rule out congenital heart disease (CHD). We should also remember some heart disease might not have specific symptoms and so it is important that we should get our child checked by pediatrician who can examine to find some clues for heart disease and will guide you and sort out your anxiety. The red-flag signs are:

- **Feeding issues:** If baby sucks for some time and takes rest, then again sucks (suck-rest-suck cycle) and taking long to feed (>20 min), or if there is increased work of breathing during feeds and increased sweating during feeds.
- **Poor weight gain:** Your pediatrician is not satisfied with weight gain. You need to weigh baby regularly. Growth charts given in vaccination charts are good to assess the pattern of weight gain.

- *Persistent rapid breathing (Rapid respiration, even during sleep):* Chest retractions, alae nasi moving though he is not having fever or cough.
- *Persistent high heart rate (even during sleep):* If you keep watching the chest and keep your hand on the left side of the chest, you will learn to feel pulsations. A mother is the best and most often the first person to notice that the heartbeat of baby is running very fast.
- *Precordial bulge:* Sometime chest is bulging on one side.
- *Pale, mottled skin:* Skin color will not be looking healthy but pale and peripheries always remaining cold.
- *Blue babies:* Baby will have blue lips, tongue and nails or turns blue on crying or effort. On pulse oximeter, $\text{SpO}_2 < 95\%$ (**Fig. 2A**).
- *Blue spell or Tet spell:* Mother gives history that baby cries without any reason many a times after waking up in the morning, takes deep breaths, gets tired, exhausted, lethargic for some time and then after some time regains normal activity. During this period, the lips and nails turn deep blue. This is a life-threatening episode and you must consult a doctor at the earliest preferably a cardiologist.
- Becomes *tired easily* and starts fast breathing.
- *Persistent cry of baby:* Baby is unhappy, not content and it is difficult to console him or her; has shrill cry but cannot sustain it (**Figs. 2B and C**).
- If your child faints or becomes unconscious or if there is he or she should be evaluated by doctor.
- If your child suffers from *recurrent pneumonia*, there may be a chance that he has a heart defect, so should consult a cardiologist.



Figs. 2A to C: (A) Sick-looking crying baby, she is not normal also she seems to be dehydrated. She needs to be investigated; (B) Blue arrow showing retraction of chest and white arrow shows flaring of ala nasi; (C) A close-up of same baby suggests bluishness in circumoral area. These babies must be investigated both for heart and lung diseases.

Q2

My daughter is 2 years old. When I took her to a doctor, he suspected congenital heart disease and advised echocardiography.

- (a) What is congenital heart disease?**
- (b) Will echocardiography confirm the doubt? Will it be harmful for her?**

Congenital heart diseases (CHD) are structural defects of the heart present since birth. Your pediatrician can recognize the presence of a CHD based on your complaints and clinical examinations, ECG, and X-ray chest. We should know that even if the disease is congenital it may not have any manifestation just after birth or for some time period.

Echocardiography is a kind of ultrasound of heart which gives exact structure and function of the heart, i.e., based on these information doctor may decide about the management plan. The two-dimensional (2D) echo/M-mode/spectral Doppler are the integrated modes of ultrasound for comprehensive evaluation of the heart.

Some CHDs need early or urgent intervention otherwise survival is at stake or irreversible damage happens. Some CHD needs regular follow-up and may be follow-up of echoes needed every few weeks. Depending on the type of CHD, decision of intervention would be taken. A serious heart defect requiring intervention can be missed up to 10–30% cases. Echocardiography is a safe procedure and has no hazards or side effects; even echocardiography is done in fetus known as fetal echocardiography (that is echocardiography of baby within mother's womb) also to know about heart defects. So, do not worry.

Q3

Can a baby with congenital heart disease lead a normal life? What are the precautions I should take for my baby?

Yes, this is a very important concern as a parent and it is you the parents who play the chief role in taking proper care of the kid. If CHD is diagnosed on time and advice given by treating doctor followed strictly most of the CHDs are curable. Many children may need intervention early or late. Majority of CHD are simple defects and those children who get treatment on time, can live up to their full potential. However, approximately one-thirds of children with CHD may not achieve full capacity and would be advised a sedentary lifestyle. A few of them may need multiple procedures.

Therefore, following precautions, we would recommend:

- To get an advice on diagnosis and management by an expert of the field.
- Please follow the advice about dietary management, medical management, or intervention strictly.
- Avoid the close contact with a person with infection (one with cough and cold, diarrhea). Caretakers must maintain strict hand hygiene. Dental hygiene of older children must be looked after. It is essential to be under the supervision of the pediatrician and prompt treatment of infection.
- If there is fever $>100^{\circ}\text{F}$ or certain intervention or surgery to be carried out, antibiotic prophylaxis needs to be given.
- Exercise for these children must be customized but must not be stopped.
- Children with a CHD are vulnerable to infections. They tend to catch infections especially respiratory infections. These infections flare up to pneumonia easily. The steps that need to be taken to protect them are:
 - Immunization to be completed on time.
 - Additional immunizations such as pneumococcal vaccine, meningococcal vaccine, and influenza vaccine need to be given.

Q4

My neighbor's son has been diagnosed to have a serious heart disease at the age of 3 months though he was absolutely normal at birth. Why were they not told that he has a heart disease at birth?

Sometimes the common heart diseases, such as "hole-in-heart" cannot be detected in first few weeks of life due to absence of murmuring sounds and absence of features such as feeding difficulty, poor weight gain, etc. These symptoms suggest heart failure (HF), which is different from the HF of older people. Most common heart diseases such as ventricular septal defect and patent ductus arteriosus may not show themselves up in first 1–4 weeks. This is because of the delay in physiological maturation of lungs. Ventricular septal defect and patent ductus arteriosus (VSD and PDA) increase the lung pressures and blood flow. They also increase the heart size. A large hole in heart creates an imbalance of blood flow between lungs and body, i.e., lungs are overflooded. A large hole in heart presents with increased breathing efforts, increased heart rate all due to the activated compensatory mechanisms. It results into slow body growth, poor or no weight gain. Symptoms commence after 4–6 weeks of life.

Atrial septal defect (ASD) is another type of hole, which may remain hidden for many years, up to adulthood. Stenosis or narrowing of the valves again can present at any age even in adults.

Q5

My pediatrician says that my baby is a blue baby. I only appreciate blueness of lips, palm, and soles, while he cries. What exactly a blue baby is? How should I care for my baby? Is there a future for these blue babies?

Congenital heart disease may cause change in color of skin, nails, tongue, and lips. This happens when the good or oxygen rich red blood mixes with the oxygen depleted blue blood and circulate in the body which becomes evident in skin, nails, tongue, and lips either always or at least when he cries. These babies have a severe heart disease which would require open heart surgery early. Few of these, babies must undergo heart surgery in first few weeks of the life.

However, pediatricians use special tool (pulse oximeter) to recognize the blueness or cyanosis. Many times, even they cannot see blueness because of color of skin or anemia of baby and borderline saturation. If pulse oximeter is showing value of SpO_2 85–95%, then it cannot be perceived visually. SpO_2 must not be done on cold hands and feet. Also, proper size probe is essential.

These babies must be monitored closely. All the precautions which we take for a child with heart disease are to be obeyed. Blue babies do not tolerate dehydration and anemia, so many of these blue babies become serious after crying which is termed as spell when they breathe fast and more blue, then lethargic—so we have to be careful if such situation arises, they need to consult a doctor immediately. They may need early operation/intervention. They may also need more than one operation. They can be advised some conservative and medical management to minimize the symptoms.

There have also been medical advances resulting in these patients living longer, fuller, and more active lifestyles. As these babies grow, they can attend and thrive in school and live an active lifestyle. They often participate in the same activities as their peers. Many of these babies with CHD grow up to be successful adults, going into careers in medicine, business and even sports [Olympian Shaun White has Tetralogy of Fallot (heart disease)].

So if your child has just been diagnosed with CHD, you should be preparing yourself to be strong enough to give the extra care and support to your baby, always connected with pediatrician and pediatric cardiologist. The child's life should have a healthy balance and moral boost to bear all the stress.

Q6

My son is very active but during school routine screening suspected that he might be having some form of congenital heart disease which came out to be narrowing of a valve and need restriction of outdoor sports. I am very worried. How will I be able to manage my child? Doctor also talked about ballooning of valve, how does it help?

Narrowing of any valve reduces the capacity to work. These children may have tiredness on running. Sometimes they may lose consciousness. A child with these kinds of complaints must undergo intervention. Usually, valves can be treated without the open-heart procedure. These ballooning procedures are safe and done in catheter laboratory. Your child would need only 1–2 days of hospital stay. Ballooning decreases the degree of narrowing, and hence the risk of syncope becomes less, and the exercise capacity improves. However, if he is not having any symptoms and echocardiography shows mild form of disease, sports can be allowed but regular echocardiography evaluation must be done. These children must have good dental hygiene and prompt and adequate treatment of the infection. They would need some antibiotics before the certain invasive procedures.

Q7

My baby is only 2 weeks old, and has been diagnosed to have congenital heart disease which will need surgery. I am very scared of surgery. Is there any medical treatment also available? What will be the quality of his life if surgery is done?

The outcome of the neonatal surgery is getting better over the years. So please do not worry. It is obvious that baby has risk to the life if left unoperated. Medical management is usually not helpful in CHDs as these are structural defect of heart which has to be repaired. However, medical management may reduce the discomfort of the baby only before the optimum time of surgery. In critical diseases, early, timely intervention brings out best results. Risk of procedure generally is <10%. Quality of life remains good if surgery has been done on time.

Children with mild CHDs that do not need surgery usually have no restrictions on their activity. However, before participating in competitive sports, a consultation with a pediatric cardiologist is required. The pediatric cardiologist will carry out an evaluation of the child and will advise accordingly.

The children with severe CHDs tend to restrict their activity as per their capability. However, supervised playing with attention to adequate hydration is encouraged. Participation in competitive sports is discouraged.

A child with CHD is encouraged to go to school and carry out normal activities. Any restriction in this respect will be detrimental for the child's self-esteem and confidence. However, it is advisable to intimate and discuss with the school teacher as to the exact nature of the child's heart problem and the limitations that need to be exercised in the school activities. It is necessary that excessive limitations are not placed on the child otherwise they tend to have a feeling of "being left out" and develop problems of social interaction in future life.

Q8

My 1-year-old baby has been diagnosed to have a hole in heart for which a device closure or surgery has been recommended. What should I do? If I go for a device how safe is it?

Device closure for atrial septal defects is done about 3 years of age. Patent ductus arteriosus are usually closed around the 1 year of age. Device closure of simple holes are preferred therapeutic procedure globally, now. The overall outcome is great because there are no surgical scars, reduced hospitalization, and minimum precautions. Nevertheless, all defects cannot be closed by devices and back-up consent for surgery is always mandated before going for a procedure. One must understand the procedure and cosmetics of procedure must not be the criteria for selection of the option. Devices are safe and they work like a brick in a broken wall. Cementing is done by the body itself. Though devices get some kind of covering within 3 days, but this covering takes around 3 weeks to 6 months to become strong and covered by body tissues. Therefore, care must be taken to avoid trauma to the chest, and to avoid infection till that period. Sometimes a blood thinner can be given for 6 months.

Q9

My first child, now 2 months old, is diagnosed to have some complex heart disease and will need multiple surgeries as advised by doctor. Is there any alternative of these surgeries for him? What all activities he will be able to do if all surgeries are successful?

When should I plan for a second baby, is there a chance that my second baby will also have CHD?

The complex CHDs demand a lot of commitment from both, parents and doctors. Surgical options are limited. Usually, a combination of catheter procedures and open-heart surgery would be offered time to time. Yet a completely normal heart may not be achieved. Overall, success rate in these cases varies between 70 and 90%. Depending on the heart condition these children can have a decent life with all routine activities without constraint, but unfortunately competitive sports, professional singing, and dancing might not be easy for them. Also pregnancy and childbirth carries more risk than normal. However, with continued care and a healthy lifestyle and mental boost they have shown success in various fields including medicine, law, engineering, and even sports.

There is a chance of borderline increase in risk of recurrence of CHD in second pregnancy, average 2–5%. Therefore, your obstetrician will keep strict vigil on you. A fetal echocardiography would be recommended between 16 and 19 weeks. If a serious CHD is diagnosed, then termination can be advised. A genetic counseling is also recommended.

Q10

My 10 days old baby suddenly became sick and the doctor said that he had coarctation of aorta. I do not want surgery for my small baby. My friend's son had aortic stenosis and underwent ballooning at the age of 4 days. He is doing well. I also want ballooning only.

What must I do?

Ballooning procedure for coarctation of the aorta is feasible and often done when a baby is >5–6 months old. Neonates have tendency to develop coarctation again and again. Therefore, surgery is preferred in these babies and has <10% chance of recurrence. Ballooning is done in very sick neonates as the rescue procedure and in babies with discrete narrowing of vessel (aorta) after 6 months of age.

Ballooning for valve (aortic or pulmonary) is different from the vascular (aortic) obstruction. Ballooning of obstructed aortic or pulmonary valve (aortic or pulmonary stenosis) of a critically sick baby is the standard recommendation across the world. Surgery in these babies born with critical aortic or pulmonary stenosis has high surgical risk involved with cardiopulmonary bypass and absence of appropriate artificial valves, and hence ballooning is preferred in most centers.