Frédéric Dallaire
Anne Fournier
Claudia Renaud

Young at Heart

Understanding childhood heart diseases

3rd edition

fondation en cœur

Young at Heart

Understanding childhood heart diseases

3rd edition

Frédéric Dallaire
Anne Fournier
Claudia Renaud

Young at Heart

Understanding childhood heart diseases

3rd edition

fondation en cœur

Coordination: Sylvie Gérin-Lajoie

Heart concept on the cover:
UNI UNITÉ CRÉATIVE INC.

Pictures: Mélissa Vincelli

Many thanks to all the photo shoots participants as well as their parents.

Illustrations: Madeleine Leduc

X-rays of Figure 10: CHU Sainte-Justine
Pictures of Figures 29 and 30: Medtronic

The translation of this edition was provided by Franglish Translations

En Cœur Foundation
8535 boul. St-Laurent, Suite 310
Montreal, Quebec
H2P 2M9
Telephone: 514-737-0804, extension 2

Toll free: 1-800-EN-CŒUR
Website: http://en-cœur.org

Printed in Canada

Dedication

This book is dedicated to children with heart disease and their parents, as well as all those who help them.

Thank you to all parents, teachers, volunteers, health professionals for caring for children with heart disease.

Acknowledgements

Several people have contributed to this book. We would like to warmly thank all the volunteers who reread and commented on the pages of this book:

› Claudia Massé
› Danielle Vachon
› Daniel Marceau
› Sandra Filopoulos
› Michele Zegray
› Virginie Carrier

We would also like to thank Dr. Christine Houde and Dr. Marc Bellavance for contributing to specific sections of this book. Special thanks to Heidi Shapiro for her important contribution to the section “Reactions to the diagnosis” and to Ginette Matte for having transcribed the previous edition in electronic format.

In addition, we are infinitely grateful to everyone who contributed to the previous editions of this book, in particular:

› Dr. Marie Béland
› Dr. Adrian Dancea
› Dr. Joaquim Miró
› Dr. André Davignon
› Denise Pépin
› Adeline Denis

Finally, we also wish to thank Sylvie Gérin-Lajoie, Executive Director of the En Cœur Foundation, for her support throughout the work involved in making this 3rd edition possible.

The translation of this edition was possible thanks to the generosity of Filo Import Inc.

Table of contents

Preface

The heart is vital. When parents learn that their child has a heart disease, they have good reason to worry. Since I have started caring for and accompanying children with heart disease, I often observe that the unknown is the greatest source of concern. “How does a heart with three valves instead of four work? What difference does it make to have a big hole between your two ventricles? What will happen at birth if the great vessels are inverted?” I see how relieved parents are to know a little more about how the heart works. I can feel how soothing it is for them to get a better idea of what life is like for a child with a heart defect.

The purpose of this book is to explain the varied heart problems and treatments available in order to alleviate worry. It is intended for families with a child with heart disease, as well as all those who want to better understand the hearts of children, whether they are healthy or have small defects.

We have changed the title, but this is the 3rd edition of *Why my child?* Hearts have not changed much, but our understanding of heart disease improved and so treatments evolved. We felt it was important to provide an updated version. Therefore, Anne Fournier, Claudia Renaud and I undertook to draft this new edition.

The heart is a fascinating organ. I never tire of studying, curing and talking about it. I sincerely hope that when reading these pages, some parents will be able to say, “Phew! It makes me feel good to have a better understanding!” Our mission will then be accomplished.

Frédéric Dallaire, Pediatric Cardiologist

Chapter 1

The heart

The normal heart and how it functions

The heart is a vital organ, but also a symbol. It is commonly believed that to show some heart is to be moved by noble sentiments: courage, generosity, love. Since antiquity, we consider it to be the very seat of life. We can therefore understand why any heart problem can be distressing, especially when it involves a child. Beyond the symbol, the role of the heart is to allow blood to circulate throughout the body. The blood carries oxygen and nutrients to various organs. It then rids them of carbon dioxide and their waste. It is the heart that first pumps blood to the lungs and then gets it back and sends it to each of our cells. The functioning of the heart as a pump is relatively simple. To understand it clearly, we simply follow the course of the blood step by step, as shown in Figure 1.

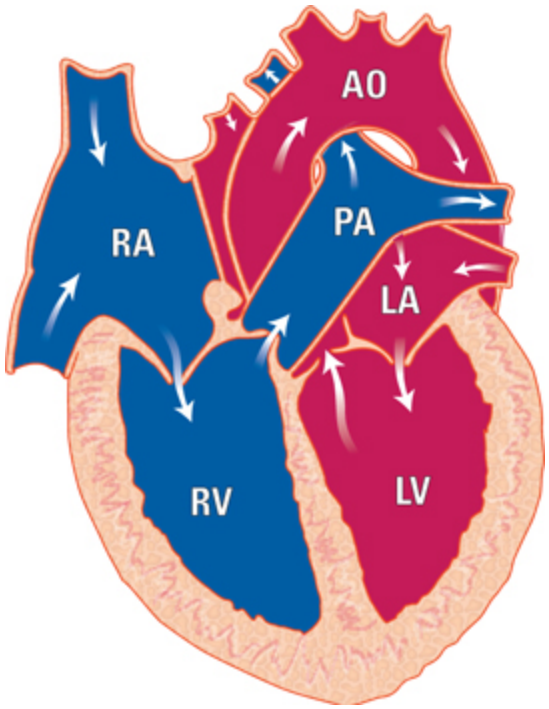


Figure 1 | The normal heart

The functioning of the heart as a pump

The heart is made of two pumps that each have two distinct chambers. The upper chamber is called the **atrium,** and the lower chamber is called the **ventricle.** The atrium serves as a reservoir for the return of blood while the ventricle pushes blood out of the heart.

On the right side, the right atrium (RA) receives oxygen-poor blood—often described as “blue” blood—from the body. The blood then travels down the right ventricle (RV) to be pumped to the lungs. This is where it gets its oxygen supply and offloads carbon dioxide gas. It then returns from the lungs to the left side of the heart.

On the left side, the left atrium (LA) receives oxygenated blood—red blood—from the lungs. The blood then travels to the left ventricle (LV) which will pump it to all of the body’s organs.

Various organs, such as muscles, liver, brain, and kidneys, will take in oxygen and nutrients. The blood, now oxygen-poor, returns to the right side

of the heart to start this same loop over and over again.

To prevent oxygenated and deoxygenated blood from mixing, the right and left sides of the heart are divided by a wall called a septum.

The heart valves

The heart comprises four valves. The valves open so blood can travel from one structure to another. They then close to prevent the blood from going back.

The **tricuspid valve** is located between the right atrium and the right ventricle. The **pulmonary valve** separates the right ventricle from the pulmonary artery (PA). The **mitral valve** allows blood to circulate between the left atrium and the left ventricle. Finally, the **aortic valve** is located between the left ventricle and the aorta (AO).

The circulation of blood in the heart and body

The ventricles are attached to large arteries that deliver blood to the organs. The atria are connected to large veins that bring the blood back to the heart.

The inferior vena cava and the superior vena cava are two large veins whose role is to transport deoxygenated blood (the “blue” blood from the body) to the right atrium. The blood then travels through the tricuspid valve and ends up in the right ventricle. After the passage of blood, the valve closes to prevent it from returning to the atrium.

Once the right ventricle is full, it contracts to pump blood to the lungs. The pulmonary valve then opens to let the blood move forward. When the contraction of the ventricle is complete, the valve closes, preventing blood from returning to the ventricle. The pulmonary artery is divided in two, one branch towards the right lung and another towards the left lung, then into several smaller and smaller branches, until they become capillaries. These vessels, thinner than hair, are in contact with the alveoli of the lungs, which

contain the air we breathe. Once in the lungs, the blood sheds carbon dioxide and absorbs oxygen.

The newly oxygenated blood returns from the lungs through the pulmonary veins (the only veins that carry oxygenated blood throughout our bodies) in direction of the left atrium. It then travels to the left ventricle through the mitral valve. Again, the valve opens as blood moves forwards and closes to prevent it from moving backwards. The left ventricle, which is the heart's most muscular chamber, contracts with enough force to allow blood to flow throughout the body, all the way to the tips of the toes. Blood leaves the left ventricle, enters the aortic valve, and travels to the aorta, which is the largest artery in the body. It is the aorta that conducts blood to all other blood vessels and, ultimately, to every cell in our body.

The aorta is divided into several branches, comprised of arteries, arterioles and capillaries, in order to distribute blood to all parts of the body. Organs and cells then exchange carbon dioxide for oxygen. The blood, now blue again, travels back to the heart through veins, returning to the right atrium through the inferior and superior vena cava. The same process is repeated over one hundred thousand times a day!

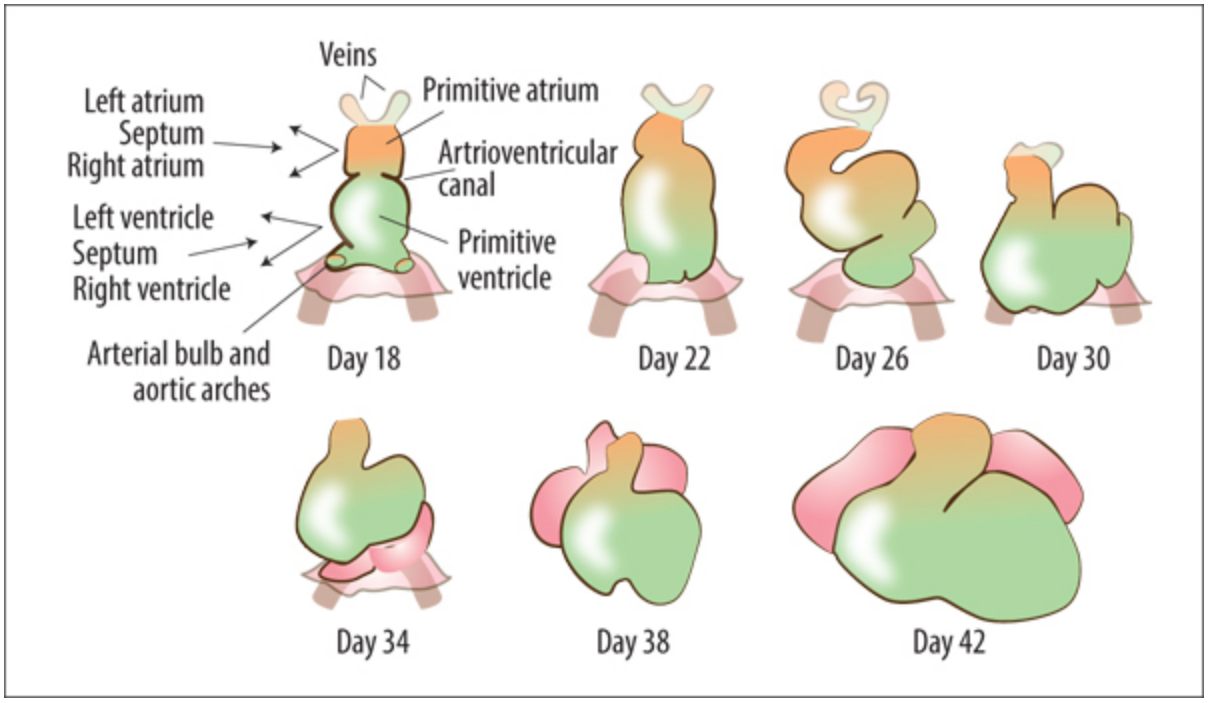
The control of the cardiac system

Each contraction of the heart begins with an electric current that travels up and down the heart. This electrical impulse originates in a small part of the right atrium called the sinus node, often referred to as the heart's natural pacemaker. The sinus node determines when the heart should speed up and slow down. The current then travels throughout the entire heart, causing the atria and ventricles to contract and then relax. It is this contraction that allows the ventricles to pump blood. The beating of our heart is the result of these rhythmic contractions. This electrical system is essential for the proper functioning of the heart.

The development of the heart

The heart develops from a small group of specialized cells located in the upper part of the embryo's chest at the very beginning of the pregnancy (Figure 2). These cells rapidly form a tube that folds back upon itself in the shape of an "S". After a few days, the tube develops bulges that will form the different chambers of the heart and the origin of the great arteries.

Figure 2 | The development of the heart (embryology)



During the first stages of the development of the heart, a primitive cardiac tube folds back on itself and changes its internal structure to form the heart.

Between these bulges are narrowings from which the heart valves will develop. Walls will soon separate the left and right side of the heart; they develop in three places: between the two atria, between the two ventricles and between the two main arteries, the aorta and the pulmonary artery.

Connections are simultaneously established between the heart and the blood vessels that are developing throughout the body. Vessels from the lungs also connect to the primitive heart. By the 10th week of pregnancy, the heart is almost completely formed. Subsequently, the heart and blood vessels continue to develop in proportion to the requirements of the growing fetus.

The circulation before birth

Before birth, it is the mother's placenta that supplies oxygen and nutrients to the fetus. The lungs of the fetus are not inflated and need only a small supply of blood to grow. Special fetal pathways in the circulation allow the blood to circulate with maximum efficiency. The blood arrives from the placenta loaded with nutrients and oxygen and then returns to the placenta through the arteries to get rid of carbon dioxide and other substances released by the tissues of the fetus.

Because the circulation before birth is complex, it is easier to follow the circulation of blood in a drawing (Figure 3). The oxygenated blood returns from the placenta to the inferior vena cava of the fetus by way of the umbilical vein, crossing the liver through a special channel called the ductus venosus. It is then directed through an opening (foramen ovale) located between the two atria. This oxygenated blood is then pumped through the left ventricle into the aorta. From there the blood goes to the developing brain and, to a lesser extent, the rest of the body. The deoxygenated blood that returns from the upper part of the body along with a small amount of oxygenated blood coming from the inferior vena cava is directed to the right ventricle which subsequently pumps it into the pulmonary artery. Only a small part goes to the lungs while the greater part of it, flowing by way of an additional conduit (the ductus arteriosus), travels through the descending aorta where it supplies the lower part of the body.

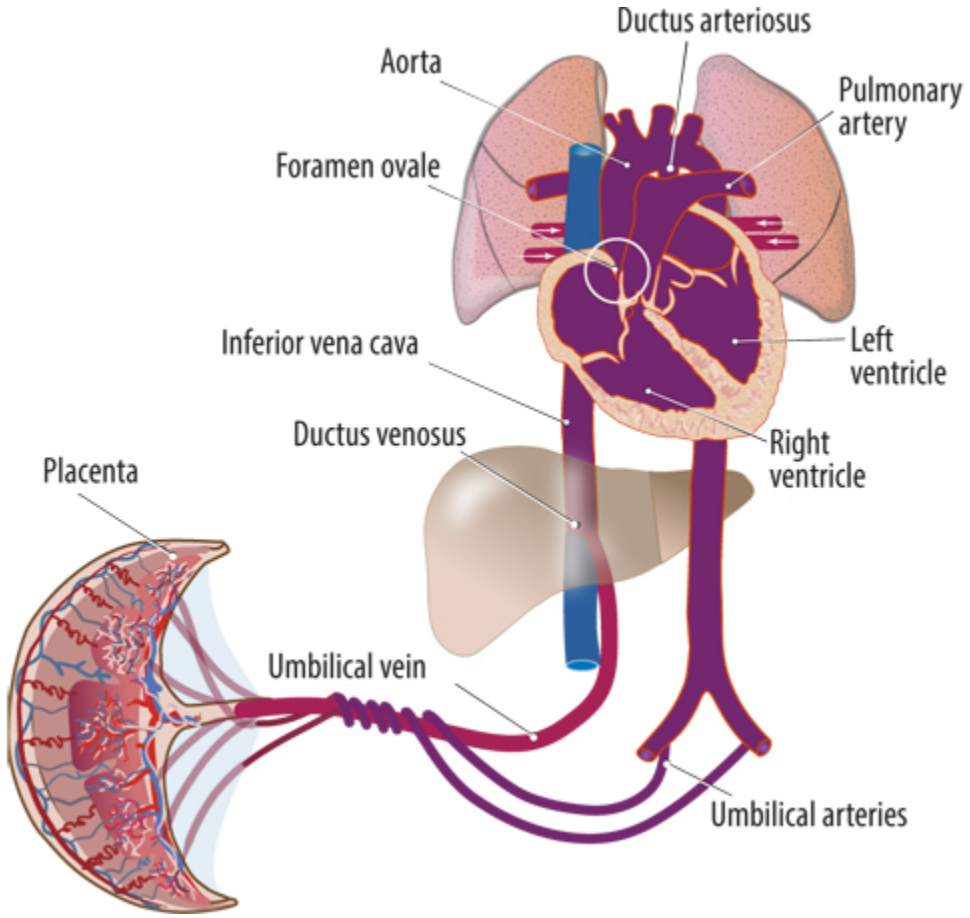


Figure 3 | The fetal circulation

Most of the blood that has become deoxygenated then returns to the placenta to be oxygenated again. Thanks to these connections and the mother's placenta, most fetuses, even those with severe heart problems, will have a normal growth in the uterus and even appear perfectly normal for the first few hours after birth.

Changes in blood circulation at birth and during the first days of life

Immediately after birth, connections to the placenta cease and the ductus venosus closes. From the first cries of the baby, the lungs swell, and blood begins to course through them in increasing quantities. The blood now takes its oxygen from the air passing through the newborn's lungs. As more blood returns to the left atrium, the foramen ovale valve closes. Over the next few

days, the ductus arteriosus also gradually closes. All of these changes dramatically increase the amount of oxygen available to the newborn after the lungs start functioning. Blood pressure in the aorta rises, and over the course of the next few weeks, the pressure in the vessels of the lung and in the right ventricle decreases. This is often when many babies with heart defects show the first signs of their disease.

Chapter 2

What is wrong

Prevalence and causes of heart defects

One child in a hundred is born with a heart defect. The majority of affected children will have little or no problems with this defect and can lead normal lives without any intervention. However, some defects are more serious. About one in six children with a heart defect will need heart surgery or catheter intervention. Fortunately, several defects can be completely corrected. However, the most serious defects, such as hypoplastic left heart syndrome, cannot be entirely repaired. With the advancement of technology, many children with severe defects can hope for a full life. Unfortunately, even today, despite all the progress in surgical techniques and medical care, some children will not survive. The risk of dying from a heart problem is still very low: it is less than 1% for all defects combined.

The causes of heart defects are numerous, but they are only known in a limited number of cases. While it is clear that genes and chromosomes are often involved, our current scientific knowledge can only explain a small portion of heart conditions. The defects may be hereditary or genetic, e.g., linked to a syndrome such as Down's syndrome, or the consequence of a disease in the mother—such as diabetes—or of taking certain medications during pregnancy. However, in the majority of cases, it is not known why a child is born with a heart defect. The risk of having a child with a heart problem has not changed much over the years. What has changed are treatments and survival: children who were once dying soon after birth (they were called "blue babies") are now living well into adulthood.

Signs and symptoms

Many heart defects are detected before a child shows any symptoms. In many cases, the family doctor or the pediatrician detects certain signs during a routine examination and refers the child to a pediatric cardiologist. In other cases, heart defects are detected as early as during the second trimester of pregnancy. Although most children with heart defects do not have symptoms initially, some may have them from birth. These symptoms can manifest themselves in different ways. In this chapter, we describe the most common signs and symptoms.

Heart murmurs in children

A murmur is simply a sound that is added to the usual heart sounds. They are the sounds a doctor hears when examining a patient with a stethoscope. Fortunately, the vast majority of heart murmurs are benign. We then say that they are “functional” or “physiologic.” They represent the normal sound of blood passing through large vessels near the heart. This type of murmur may be heard in over a third of healthy children. Since this is a normal heart sound, having a functional murmur does not require any treatment or follow-up. In other words, a heart murmur is not synonymous with heart defects.

However, some defects may also explain the presence of a heart murmur. We then say that the murmur is “pathological” or “organic.” The sound may be caused by blood passing through a constricted heart valve or by abnormal connections within the heart. The intensity of the murmur does not necessarily reflect the severity of the abnormality since some mild defects can cause a murmur that is quite loud and easy to hear.

The characteristics of the murmur, in particular its sound and intensity, generally make it possible to distinguish a functional (normal) murmur from a pathological (abnormal) murmur. Sometimes, additional tests are necessary to determine the nature of the murmur (electrocardiogram,

echocardiogram, X-ray of the heart, etc.). These tests are described in Chapter 3.

The intensity of murmurs changes over time. Most of them, especially the benign ones, decrease in intensity when the child is calm or asleep. They become louder after physical exertion or when the child is anxious, feverish or anemic. Many functional murmurs appear around the age of two and go away by puberty.

Palpitations and arrhythmia

A palpitation is a sensation that the heart is beating irregularly or too fast. Often, the child will describe chest pains, a sensation of skipped beats, or will speak of having the impression that his heart is trying to come out of his chest or even of his neck. The palpitations may be due to heart disease, but most of the time, they are mild and do not require special treatment. Someone with no heart problems may experience palpitations after an intense emotion, stress, a physical effort, or even without any apparent reason. Palpitations may also be caused by an arrhythmia. An arrhythmia is an episode during which the rhythm of the heart becomes abnormal (too fast, too slow, or irregular).

Not all palpitations are arrhythmias. In addition, arrhythmias are not necessarily dangerous. It is up to the physician to determine the cause of the palpitations based on the symptoms and, if necessary, to request additional medical examinations. In children with a heart defect, it is important to see a physician if new palpitations occur, and to do so quickly if they persist for several minutes or if they are accompanied by symptoms such as dizziness, pallor, shortness of breath or loss of consciousness.

In children as in adults, the heart beats regularly at a rate that varies according to age or the level of activity. The hearth rhythm is the result of a series of well-defined electrical events that occur in specialized heart muscle cells. A small group of these cells located in the right atrium (sinus node) regularly emit an electric wave (nerve impulse) which spreads throughout both atria causing them to contract. A few hundredths of a

second later, the impulse reaches another mass of nerve cells further down in the right atrium and from there the impulse is conducted to the ventricles through specialized nerves. This is how the heart beats: the contraction begins in the atria, then moves down to the ventricles. This sequence occurs on average almost 100 times per minute in children, or about 150,000 times per day.

Sometimes the impulse is transmitted poorly and irregularly. The heart may then beat very quickly (tachycardia), very slowly (brady-cardia) or irregularly. Arrhythmias are usually benign. They are only of concern if they cause discomfort for the child or impairs the functioning of the heart by causing the heart to contract too quickly or too slowly.

Various medications may be used to treat arrhythmias. In some cases, it is also possible to use special catheters to find and eliminate abnormal nerve connections in the heart.

Heart failure

Heart failure is the inability of the heart pump to function normally, that is to say to supply the body with necessary blood for it to work properly. Organs that do not receive enough blood will have reduced or abnormal function. For example, the muscles of the body will not be able to work normally, and the child will therefore feel more tired. The newborn or infant will have trouble feeding, which will lead to insufficient growth and weight gain. It will be difficult for the liver to rid the body of waste as well as to synthesize essential building blocks such as protein. The kidneys will not be able to excrete waste from the body. Due to the failure of the heart pump, the pressure in the veins will increase and cause shortness of breath with exertion, an enlargement of the liver and water retention in the tissues (oedema). Usually, heart failure sets in slowly, and it takes several days for parents to figure out that something is wrong.

Cyanosis

Cyanosis, or a bluish colouring of the skin, remains a striking symptom, especially in the newborn. It is relatively rare in older children because of the progress of heart surgery. This abnormal colouring of the skin is due to a decrease in blood oxygenation. Normally, the blood that courses through the arteries is filled with oxygen and therefore is bright red. There are two principal causes for cyanosis: either the blood did not normally pass in the lungs, which occurs in certain heart defects (tetralogy of Fallot, tricuspid atresia, etc.), or the lungs are not functioning normally and are unable to sufficiently oxygenate the blood, which occurs in some lung diseases.

Cyanosis in infants and young children is a worrying symptom for both parents and healthcare professionals. However, it should be noted that newborns often have what is called acrocyanosis, which is cyanosis that is not caused by a problem with the heart or lungs. Indeed, during the first weeks of life, the edges of the lips, the palms of the hands and the soles of the feet of a healthy newborn baby may turn bluish. This is due to the decrease of blood flow to the extremities, which is quite normal. Furthermore, a normal newborn who cries vigorously can hold its breath long enough to show a slight cyanosis of the lips and even of the entire body. A bluish tinge around the mouth, called circumoral cyanosis, also regularly occurs when a child is cold.

When cyanosis is caused by a heart defect, the discolouration is constant. It is important to mention that although the amount of oxygen in the blood is reduced, the organs of the body, especially the brain, do not generally lack oxygen, unlike in the case of heart failure. However, children may get tired more easily than others and have stunted growth. If cyanosis persists for a while, the bone marrow will respond to a shortage of oxygen by producing large amounts of red blood cells and the blood will gradually thicken. This thickening of the blood, called polycythemia, is initially useful because it allows a greater number of red cells to carry oxygen. Over time, if cyanosis persists, the blood will become too viscous and it will be difficult for it to travel through the vessels and capillaries. Persistent cyanosis will also cause the tips of the fingers and toes to swell. This is called clubbing.

Children whose blood is more viscous than normal are very sensitive to dehydration which contributes to making the blood even thicker. They must therefore be watched very closely in the summer; exposure to very hot weather must be avoided and the child must be provided with plenty of fluids to drink. Even though the blood is thicker, clotting may be disrupted, and the child may bleed more easily and for a longer period.

Fortunately, most congenital heart defects that cause cyanosis can be completely corrected. Rarely, a complete correction is not possible, but cyanosis can be greatly improved with a series of operations that help send more blood to the lungs. It is now very rare to see a cyanotic child that medicine or surgery cannot help.

Chest pains

Chest pain, in the form of tightness or cramping, is an important symptom in adults. When the small arteries that supply the heart become blocked, the heart muscle lacks blood. The pain felt in the chest is called angina pectoris.

This type of heart pain is very rare in children. Those who have them may complain of chest pain, sometimes even on the left side at the level of the heart. However, in most cases, it is the muscles and joints of the rib cage that are the cause of the pain. The doctor will help determine if chest pain is caused by the heart.

Syncope

Syncope, a medical term meaning “loss of consciousness,” is a phenomenon caused by momentarily low blood pressure in the brain. When the pressure in the brain decreases, several symptoms may appear. You may feel dizzy, see small black dots or have blurred vision, hear a hissing sound, feel nauseous or vomit, feel very hot, cold, or sweaty, turn pale, and feel your heart beat very quickly. If the pressure does not rise, then you will lose consciousness.

Syncope is very common, especially in teenagers. In most cases, episodes are not caused by heart disease and are not dangerous. A mild syncope may be caused by dehydration, hunger, strong emotions, standing too long, seeing blood, and taking certain medications or drugs.

However, rarely, syncope may be caused by an arrhythmia (heart that suddenly beats very fast or very slowly, to the point of becoming inefficient) which is more dangerous. It is important to contact a doctor if the child with a known heart condition faints, especially if the loss of consciousness is sudden or occurs during physical exertion. The doctor will review the specific history of the loss of consciousness to establish whether it is an arrhythmia or a mild syncope. Some tests could be done to try to clarify the cause.

Respiratory infections in children with heart disease

Some children with heart disease are more likely than others to get respiratory infections. This is more likely in children with a defect that increases blood circulation in the lungs (ventricular septal defect, atrial septal defect, complete atrioventricular septal defect, patent ductus arteriosus, etc.). The engorgement of the pulmonary vessels by this overabundance of blood disturbs the usual defence mechanisms of the lungs and bronchi against infections. Respiratory infections may therefore be more frequent, last longer and also tend to be more serious.

The simplest and most effective way to prevent respiratory infections is to restrict the child’s—and especially the very young child’s—contact with other people who have a respiratory tract infection (cold, bronchitis, flu, etc.).

The recommended immunization schedule should also be followed. The cardiologist may suggest additional vaccines to prevent certain respiratory infections.

A respiratory tract infection may sometimes make a child’s heart condition worse. This is because the child needs all its lung tissue to get enough oxygen. Additionally, respiratory fatigue which is seen in some serious infections of the lungs and bronchi may increase the risk of heart failure in some patients. It is therefore important to see a doctor if the respiratory infection is accompanied with shortness of breath.

Chapter 3

How to find out what is wrong

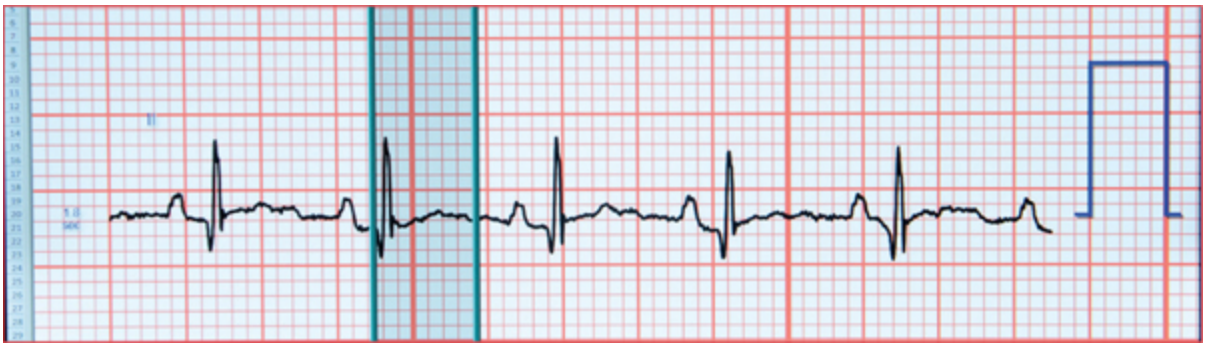
If a doctor suspects a heart defect, there are several ways to clarify the diagnosis to decide on the appropriate treatment. The necessary examinations may be quick and simple or long and complex. The most common tests in children are the electrocardiogram, Holter, cardiac event recorder, stress test, chest X-rays, echocardiogram, chest computerized tomography, cardiac magnetic resonance imaging, and cardiac catheterization.

The electrocardiogram

The electrocardiogram or ECG is easy to perform and provides valuable information about the electrical currents generated by the heart (Figure 4.1). For example, it indicates the rate at which the heart is beating (Figure 4.2), whether it is beating regularly and whether the different parts of the heart are contracting in the usual order. Measuring the strength of the electrical waves produced by the heart also provides information as to the degree of hypertrophy (or thickening) in any of the heart chambers. For instance, it is easy to tell, by looking at an electrocardiogram, if the left ventricle is abnormally thick because the main valve of the aorta (aortic valve) is too narrow and obstructs the passage of blood. The electrocardiogram will help clarify the diagnosis suspected by the doctor upon auscultation.

Figure 4.1 | Electrocardiogram

Figure 4.2 | Electrocardiographic tracing

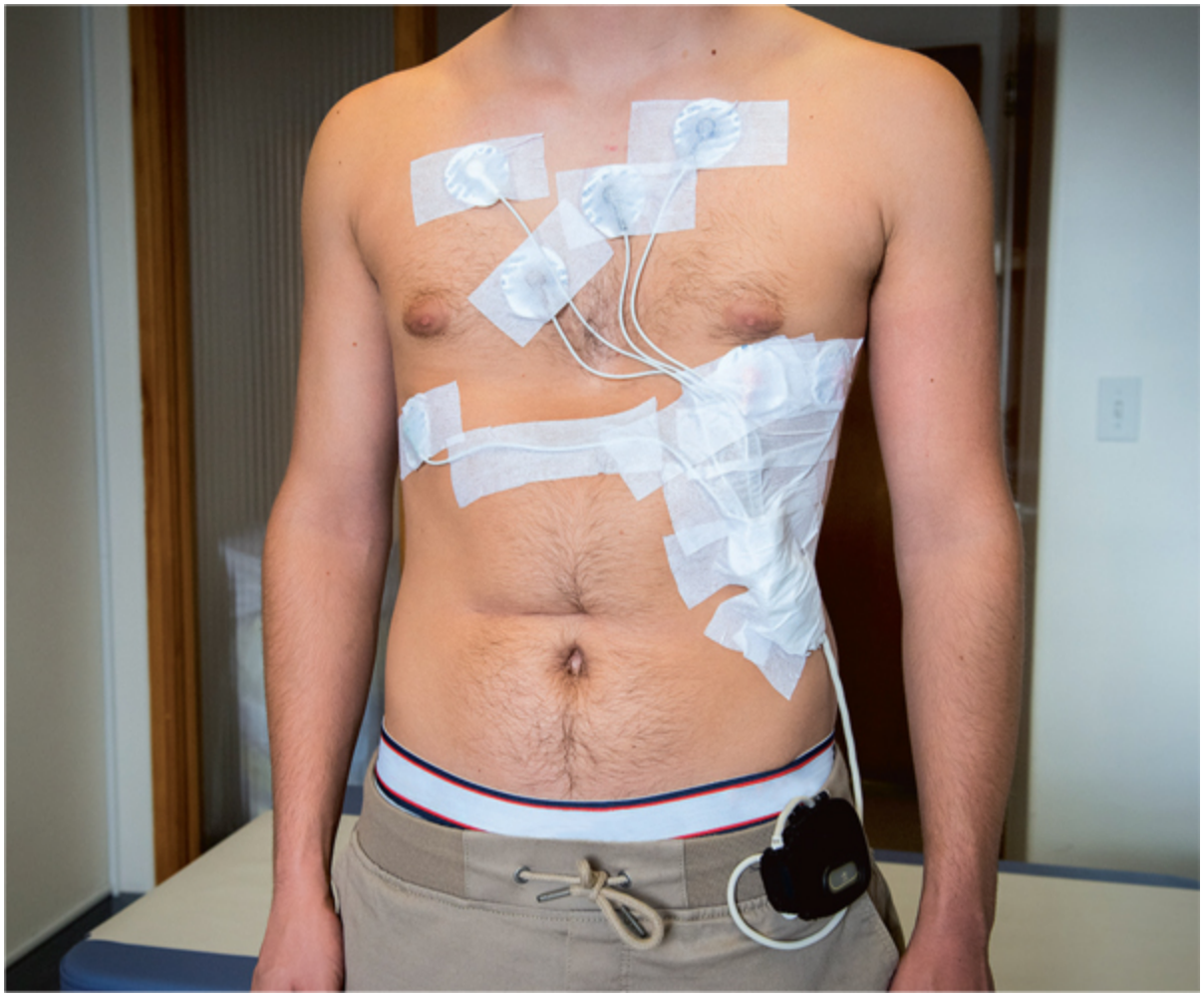


The Holter, the cardiac event recorder and the stress test

In certain circumstances when the child has an irregular rhythm, it may be necessary to assess the frequency and severity of these irregularities over a 24-hour period. The patient is then equipped with a small instrument called a **Holter** monitor (Figure 5) that records all heart beats for a full day. The

recording is then analyzed in detail. When rhythm irregularities occur infrequently and would thus be very difficult to detect in a single 24-hour recording, the patient is given a **cardiac event recorder** (Figure 6). All the patient has to do is apply this recorder over the chest when he feels a discomfort or palpitations. The heart beats are then recorded for 30 seconds. The results can be sent immediately by telephone to the hospital, where they are analyzed.

Figure 5 | Holter



A **stress test** is an electrocardiogram performed during exercise on a treadmill or on a bicycle (Figure 7). It makes it possible to detect heart muscle pain during exertion (much more useful in adults who suffer from angina), to measure the child's capacity for exertion (before or after surgery

for example) or to determine if abnormal beats (arrhythmia) are triggered by exercise.

Figure 6 | Home ECG transmitter

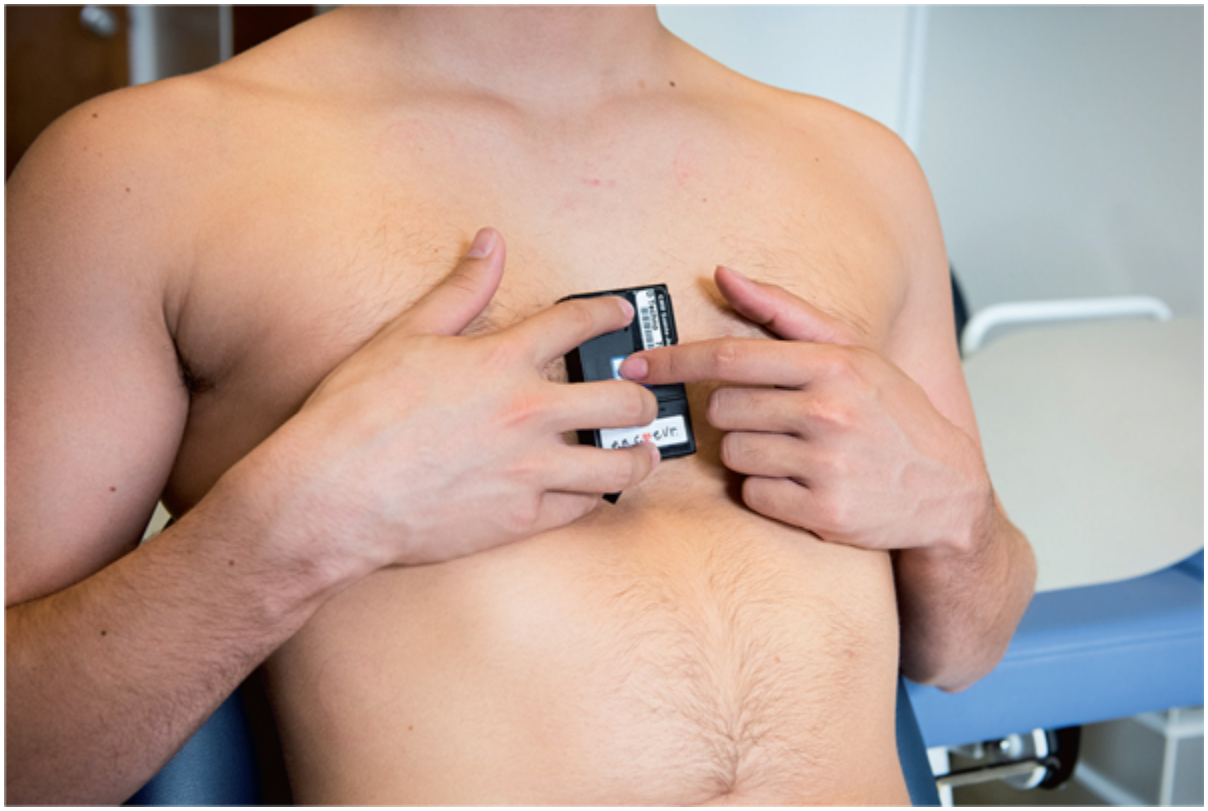


Figure 7 | Stress test

Chest X-rays, chest computerized tomography and cardiac magnetic resonance imaging

Chest X-rays used to be employed routinely in nearly all children with heart murmurs. It exposes the child to some radiation, which fortunately is very minimal. With the advent of non-radiation diagnostic methods, they have been used less frequently, and only when the total size of the heart or the flow of blood to the lungs must be evaluated.

A **chest computerized tomography,** also called a CT scan, aims to obtain images of the inside of the human body. It also uses X-rays delivered at low doses (irradiation equivalent to about 50 chest X-rays). The development of

computerized tomography techniques constitutes a new diagnostic tool with remarkable anatomical precision for cardiologists. The principle of chest computerized tomography is to obtain X-ray sections of the human body (tomography) which are reproduced as images by computer processing. This is how the vessels of the heart (aorta and pulmonary arteries) can be studied in great detail. However, the structures inside the heart (valves or walls) are more difficult to analyze because of their incessant movements.

Magnetic resonance makes it possible in a completely harmless way (without radiation) to obtain images of the heart structures in two or three dimensions, for the heart chambers as well as for the large veins and arteries. It is used as a complement to echocardiography and cardiac catheterization. This technique is based on the principle of capturing the energy emitted by hydrogen (protons) from the cells when this chemical element is subjected to variations of the electromagnetic field. The different tissues are distinguished according to their hydrogen concentration. Magnetic resonance therefore records an entire anatomical volume over a given period of time, including the dynamic events that occur. With the help of a powerful computer and software, it is possible to reconstruct a three-dimensional image of the entire anatomy. Magnetic resonance is mainly used in older children and teenagers, in whom echocardiography images are of lower quality. This technique also has limitations: the patient must be lying down in a magnet in the shape of a narrow tunnel for some time. The patient must be immobile, which requires sedation or anesthesia in infants and young children. The presence of metallic objects in the body, for example some prosthetic valves, pacemakers or epicardial stimulation electrodes, is sometimes a contraindication to magnetic resonancee.

Echocardiography

By applying an ultrasound probe on the chest, echocardiography produces an image of the heart in motion (Figure 8.1). This examination can also determine the direction of blood circulation inside the heart and measure its velocity.

Physical principles of echocardiography

This is a rather simple application of ultrasound, similar to that used with sonar during World War I for the detection of enemy submarines. A probe placed on the chest emits ultrasound which penetrates the body and returns to the probe whenever it encounters an obstacle. In the case of a living person, these obstacles are ultrasound-reflecting tissues: heart walls, interventricular septa, heart valves, etc. A computer coupled to the ultrasound-producing device makes it possible to calculate accurately the location that reflected the ultrasound. The computer then reproduces on a screen all the structures encountered by ultrasounds. A section of the heart then appears on the screen (Figure 8.2). Since the computer does this work about 30 times per second, it is possible to visualize the heart in action, that is, to see it beating on the screen. The physician may thus measure the dimensions of the different heart chambers and observe how they contract. The physician is also able to see if there is a communication between the heart chambers, valves that do not open or close normally or an anomaly in the diameter, shape or site of insertion of the large vessels of the heart.

Ultrasounds used in echocardiography are not dangerous. The energy emitted by this kind of ultrasound is much less than that of a cell phone.

Figure 8.1 | Echocardiography

Figure 8.2 | Echocardiography image of a normal heart

The Doppler

Echocardiography also measures the velocity and direction of the blood circulation within the heart. This is one of the medical applications of the Doppler effect. This phenomenon was first described over a century ago by an Austrian physicist. He had observed that when sound is reflected by a moving surface and recaptured, its frequency (number of waves emitted per second) is no longer the same. Therefore, if the object moves away from the

ultrasound emitting probe, the frequency of the reflected ultrasound captured is slightly lower. However, if the object is approaching the probe, the frequency increases slightly in proportion to the velocity of the object.

In the case of the heart, the ultrasounds are reflected by the red cells. A computer calculates the difference in frequencies, which helps determine the velocity with which red blood cells—and hence the blood in which they are contained—move away or move toward the probe. It therefore becomes possible to assess the velocity of blood flow in the vessels and in the heart. This velocity will be greater if the blood has to go through an obstacle, for example a valve that is too small (narrowing or stenosis of the pulmonary valve or of the aortic valve). It will then be proportional to the severity of the stenosis.

The colour Doppler

Colour Doppler uses the same principle to obtain an almost instantaneous view of all the velocities in the dif ferent heart chambers. Those velocities are then represented on the screen using colours: when the blood moves away from the probe, they appear in blue and when the blood approaches the probe, they appear in red. The red and the blue are lighter in colour when the velocity is higher. When the velocities are very slow, the circulation appears dark blue or red. Using this technique, it is therefore easy to identify, for example, a small opening between the left ventricle and the right ventricle (ventricular septal defect) since the blood passes at a very high speed between the two chambers and its passage is immediately visible in very light colours on the screen.

Echocardiography: a key examination

Echocardiography now provides enough information about the nature of the defect to make accurate diagnoses possible without further investigation. Formerly, almost all children needing heart surgery had to undergo cardiac catheterization and angiography. These are invasive techniques because

they require puncturing a vein or an artery. Now, the information obtained by ultrasound is sufficient in most cases.

When the images are not clear enough (as may be the case in older children and adults), transesophageal echocardiography may be used to get much closer to the heart, so that the structures such as the atrial septum and the pulmonary veins can be viewed. Ultrasound images are obtained using a very small probe that is inserted through the mouth, the end of which, once in the esophagus, allows the heart to be seen with great precision.

Fetal echocardiography

Before birth, there is a way to reliably assess whether the unborn baby has a heart defect: fetal echocardiography. This is the same technique as that used in children. Ultrasounds allow a fairly clear view of the fetal heart relatively early during pregnancy. It is thus possible to assess its functioning, detect certain heart defects and diagnose the nature of arrhythmias that may occur in the fetus.

Fetal echocardiography is prescribed in two cases: when the obstetrician suspects a problem with the fetal heart or when the risk of having a heart defect is higher. For example, a fetal echocardiogram will be ordered if the parents or siblings have a heart defect, if the growth of the fetus is too slow, if the pregnant woman suffers from certain diseases such as diabetes or if she is taking certain medications.

Ideally, it is recommended that the echocardiogram be done between the 18th and 22nd week of pregnancy. Nonetheless, when a problem with the fetal heart is suspected, the examination may be performed at any time. This is absolutely safe for the fetus: the same ultrasounds are used as for the regular obstetric ultrasound.

Most serious heart defects can be seen using fetal echocardiography. However, sometimes defects go undetected. This is the case with some smaller defects which can be very difficult to see. Others, more serious, may also go unnoticed, either because they are not easily diagnosed or

because the fetus is misplaced, making it difficult to see the heart adequately.

There are two parts to an appointment for a fetal echocardiogram: the echocardiogram itself (taking pictures of the fetal heart) and the appointment with a specialist in fetal cardiology. The images are analyzed by the cardiologist who then meets with the couple to discuss the results. Usually, there is no major heart defect and expectant parents are relieved.

When a heart problem is detected, the cardiologist will explain to the couple the nature of the defect and its impact on the child's life. Depending on the severity of the defect, joint follow-up with the high-risk pregnancy team will be offered and a birth plan will be put in place to properly prepare the care that the child may need at birth. In the case of severe heart defects, a difficult discussion about possible medical termination of pregnancy or palliative care could also take place.

Cardiac catheterization and angiocardiography

Cardiac catheterization is a specialized examination in which small tubes called catheters are inserted into large veins and arteries of the body. These catheters are then guided into the heart. They are used to measure blood pressure and blood flow accurately, as well as to take blood samples from the chambers of the heart to determine the amount of oxygen. With the help of a special dye, precise images of certain vessels that cannot be seen clearly on an echocardiogram may also be obtained. This is called **angiocardiography.** Catheters may also be used to convey prostheses and balloons to repair some defects without surgery.

Figure 9 | Cardiac catheterization laboratory

However, although echocardiography has become the main diagnostic test and has helped to significantly reduce the number of cardiac catheterizations (Figure 9), it unfortunately does not provide all the information. Catheterization is therefore some-times necessary. During this examination, it is possible to take an accurate measurement of the resistance of the pulmonary vessels to the blood flow and/or the quantity of blood shunted inside the heart through atrial or ventricular septal defects. In addition, echocardiography provides little information on certain anatomic details that are often essential before making an operative decision. Only catheterization properly allows the observation of the veins going to the heart from the lungs and the rest of the body, the coronary arteries (the small arteries that supply the heart muscle), the aorta and the pulmonary arteries. In all these cases, cardiac catheterization will be used to pinpoint the defect and safely prepare the patient for cardiac surgery, thus avoiding unpleasant surprises during the operation.

Over the years, catheterization techniques have improved considerably, and the risks have greatly decreased. In more than 98% of cases, the procedure is carried out without any inconvenience for the patient. Although they are

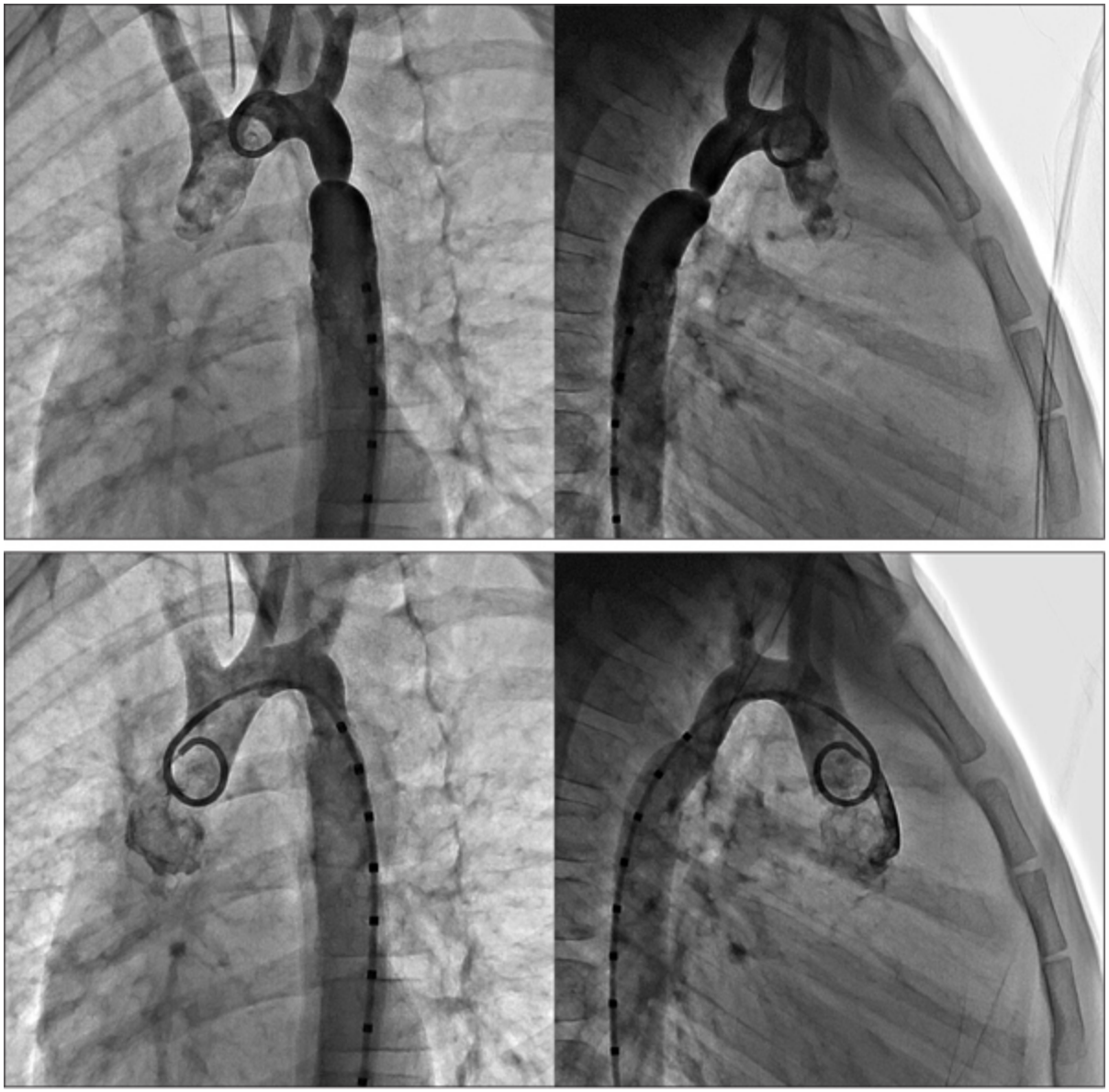
exceedingly rare, serious complications and even fatalities may occur during cardiac catheterization. The children at increased risk are those whose condition prior to the test is precarious and also certain newborn infants. Possible severe complications include perforation of myocardial walls during the manipulation of the catheter inside the heart, arrhythmia and embolization of clots or air bubbles that could then spread throughout the body. When significant blood loss occurs, a blood transfusion may become necessary. In spite of these possible complications, cardiac catheterization remains a very safe and useful diagnostic measure at any age. Whenever the cardiologist thinks the risk may be increased, he aler ts the parents and discusses the benefits and risks of the procedure with them.

Cardiac catheterization usually comprises several steps. The child is first given a sedative by intravenous or intramuscular injection that induces sleep for the greater part of the procedure and that keeps the patient from feeling pain. After local anesthesia, one or more catheters are introduced through the skin in the groin and into the femoral vein or the femoral artery. Those catheters are pushed through until the tip is guided into one of the heart chambers. As the tip is curved, it is generally easy to inspect all the chambers of the heart. When going through the femoral vein, the catheter will reach the right atrium, the right ventricle and the pulmonary artery can be reached. If the catheter is introduced into the femoral artery, it is advanced into the aorta and from there the tip of the catheter will be passed through the aortic valve and into the left ventricle. Sometimes, abnormal communications between the chambers allow access to the left heart chambers from the femoral vein or vice versa. These catheters can measure the pressure and the oxygen saturation of the blood in the chambers of the heart. Abnormalities in saturation or pressures help to assess more accurately the severity of certain defects. At the end of the procedure, the doctor removes the catheters and applies pressure to the groin vessels for about ten minutes to prevent bleeding. The patient is then returned to a recovery room for a few hours.

Angiography is done during cardiac catheterization. It involves injecting an iodine substance called a “contrast agent” that is visible through X-rays into one or the other heart chambers (Figure 10). The passage of this substance

in the chambers of the heart is recorded to be analyzed in detail frame by frame. This will make it possible to obtain precise anatomic detail of the defects, for example, the size of the heart chambers, the size of the abnormal communications, abnormalities in the distribution of the veins or arteries, etc.

Figure 10 | Angiography images (coarctation of the aorta)



Angiogram images showing coarctation of the aorta before (top images) and after balloon dilation (bottom images). The catheter used to inject the dye to better see the aorta is clearly visible in the images.

Angiocardiography adds no significant risks to cardiac catheterization itself. Contrast substances have a very low toxicity, and their iodine concentration is low. Furthermore, radiological equipment is far more sensitive and the quantities of dye that have to be injected have diminished considerably in the past few years. The principal danger is an allergy to iodine, which is extremely rare in children.

Upon discharge, the doctor carefully examines the circulation in the child's legs to make sure there is no artery or vein damaged by the procedure. It is preferable to let the child rest at home for a day or two. Parents are advised to call the hospital or take the child to the emergency room in case of doubt (unexplained fever, pain in the leg, swelling or bleeding in the groin). However, complications are very rare, so the child can resume normal activities two or three days after the operation.

The electrophysiological study

During cardiac catheterization, an electrophysiological study may be performed. This involves using special catheters that measure and record the electrical activity of the heart. By measuring how electrical impulses travel through the heart, an accurate diagnosis of an arrhythmia can be made. In many cases, treatment may even be offered for an arrhythmia that is causing symptoms or that may be dangerous. Some of these catheters are used to make an "ablation," which is a small scar inside the heart in specific places. These scars prevent electrical impulses from spreading. Thus, arrhythmia can be improved and even completely cured.

Chapter 4

The different heart diseases in children

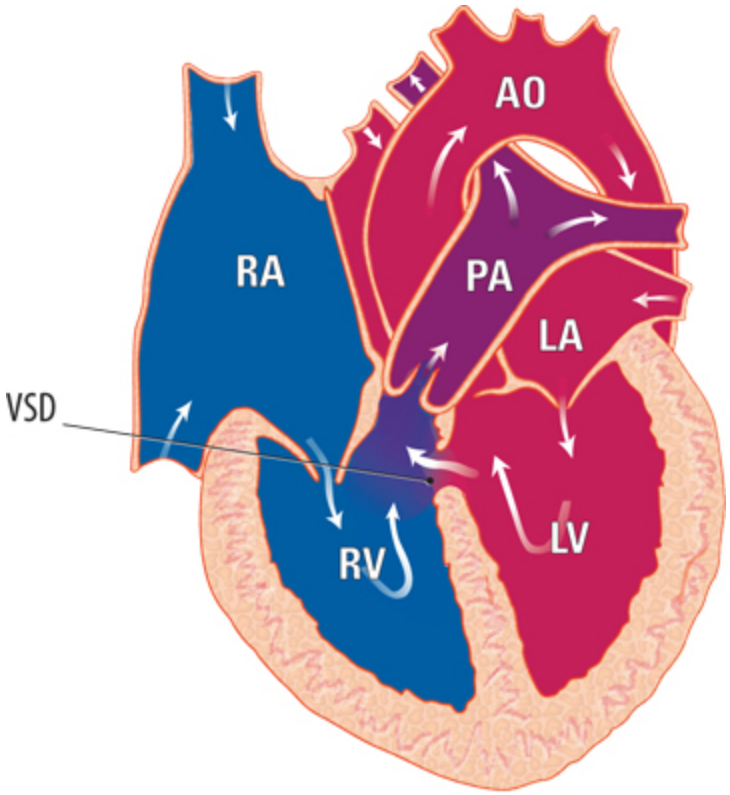
Although there are over a hundred different heart defects, only about a dozen of them account for about 90% of cases. All the others are much more uncommon. There are also some heart diseases that may appear later and affect children with normal hearts. In this chapter, we describe the most common heart defects and problems.

During the development of the fetal heart, some parts may unfortunately not develop normally (the anatomy and normal development of the heart are described in Chapter 1). The following are some possible defects in the anatomy of the heart:

- abnormal openings between the different chambers of the heart;
- a heart valve that is too narrow (stenosis) or that is leaking (insufficiency);
- one side of the heart or a heart valve may be missing;
- abnormal connections of the veins leading to the heart or arteries or arising from the heart.

All of these problems will interfere with the normal functioning of the heart.

Figure 11 | Ventricular septal defect



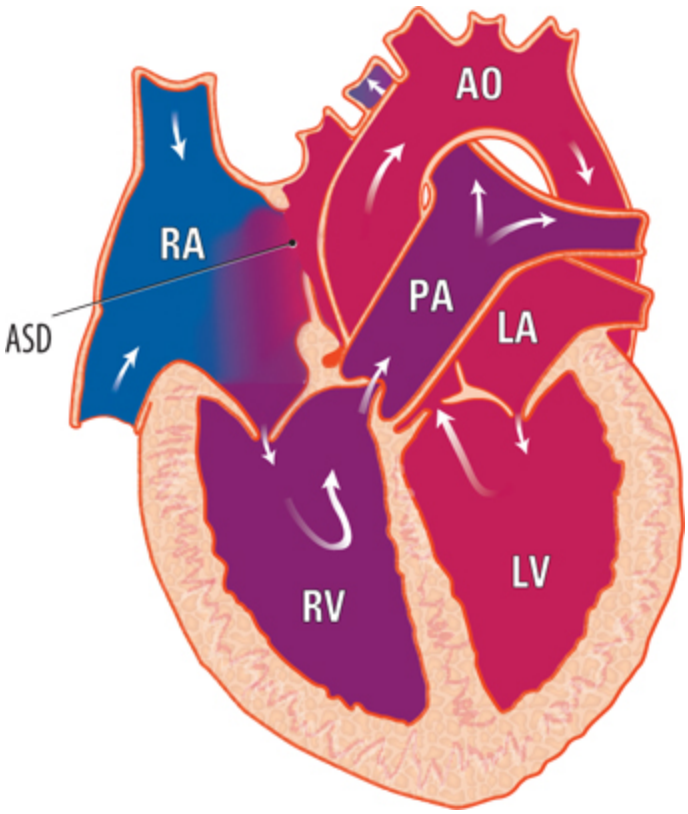
Ventricular septal defect

Ventricular septal defect (VSD) is one of the most common heart defects. It is defined by the presence of an opening in the wall that separates the left ventricle from the right ventricle (Figure 11). Because there is more pressure in the left ventricle, VSD allows for the blood to go from the left side to the right side of the heart. There is therefore well-oxygenated blood which, instead of going to the organs of the body, returns to the pulmonary artery and the lungs. If the ventricular septal defect is large, circulation overload in the lungs results. This can cause various symptoms: rapid breathing, susceptibility to respiratory tract infections, difficulty gaining weight and poorer exercise tolerance. Fortunately, most ventricular septal defects are small and do not cause symptoms or require treatment. As the child grows, ventricular septal defects tend to shrink and may even close spontaneously. Thus, even if the ventricular septal defect is quite large to begin with and the infant needs to take medication to reduce symptoms, treatments may stop over time.

However, if the defect produces symptoms that do not improve, it will have to be closed by open-heart surgery. This is usually done between three and six months of life, but depending on the type of defect and symptoms, it may be performed sooner or later.

In short, VSD is a very common heart defect and is most often benign. Even when it is larger and requires surgery, the results are usually very good, and the child can live a normal life.

Figure 12 | Atrial septal defect



Atrial septal defect

Atrial septal defect (ASD) is relatively common (10 to 12% of all heart defects) and is characterized by the presence of a hole or opening in the wall separating the two atria of the heart (Figure 12). The oxygenated blood from the left atrium therefore goes through the defect into the right atrium and needlessly returns to the lungs. If the opening is small, little blood

passes from one atrium to the other and the opening is not a problem. But the larger the opening, the more blood will flow back to the lungs, which then increases the workload of the heart and leads to various symptoms: exercise intolerance, recurrent lung infections, rapid breathing. In a very young child, there is usually little blood crossing from left to right through the opening, even if it is large. However, as the child grows, the quantity of blood returning to the lungs increases and symptoms appear or become more severe.

Atrial septal defects are usually located in the centre of the wall that separates the two atria. The newborn will often have a hole of one or two millimetres in diameter that will close spontaneously. Openings of five to six millimetres may also close spontaneously within a few months. When the openings are larger, it may be necessary to close them if they do not get smaller over time. In most cases, surgery is not required as it is possible to install a prosthesis to block the opening. This prosthesis is placed using a catheter inserted into a large vein of the groin.

In some cases, depending on the size and position of the opening, open heart surgery will be necessary. The surgeon then closes the opening, either directly with stitches or by using a piece made of synthetic material or using the part of the sac that contains the heart.

It does not matter whether the opening is closed with a prosthesis or during surgery, the procedure should be done preferably before school age, sometimes earlier if the child has significant symptoms. In general, the results are excellent, and the child can lead a normal life.

Patent ductus arteriosus

In the fetus, given that the lungs are not functional; a vessel called the ductus arteriosus connects the pulmonary artery to the aorta (see the circulation before birth section). This allows blood from the right ventricle to bypass the lungs and return to the placenta (Figure 13). At birth, the

ductus arteriosus closes fairly rapidly. In some cases, it remains open: this condition is called patent ductus arteriosus.

If the duct is relatively large, an important quantity of blood will be able to flow from the aorta to the pulmonary artery. The amount of extra blood that travels to the lungs can overload them. The child then presents with faster breathing and intolerance to feeding or exertion. If the duct is small, it usually does not cause symptoms.

If the duct is large, or is causing symptoms, it should be closed. This is usually done using a coil or a device that is inserted into the vein or artery in the groin through a catheter. This technique works well and has few complications. In rarer cases, surgery is necessary, especially in premature babies, or when the duct is too large.

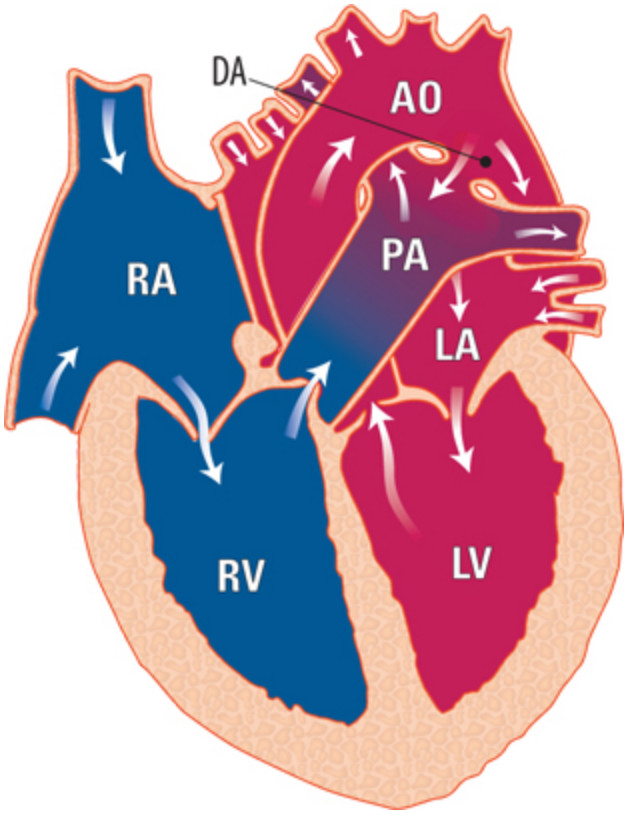


Figure 13 | Ductus arteriosus

Coarctation of the aorta

Aortic coarctation (COAO) is a narrowing (or stenosis) of the aorta. The aorta is the most important artery in the body. The narrowing is usually found just below the origin of the artery that allows the blood to flow to the left arm (left subclavian artery) (Figures 10 and 14). The severity of the coarctation may vary, but if it is severe, it will be difficult for the blood to circulate beyond the narrowing. The blood pressure in the vessels located before the coarctation will be high (arms and head), but it will be much lower in the vessels located after the narrowing (abdomen, kidney, liver, legs, etc.).

There are two typical manifestations of this defect. In the first case, the obstruction occurs in the infant and progresses rapidly in the first days or weeks of life. The rapid decrease in blood pressure and perfusion in the lower body is often poorly tolerated by the child. In addition, the heart may have difficulty pumping blood because there is too much pressure in the aorta. Those children usually need urgent surgery.

In the second case, the obstruction is less severe, and the heart has time to adjust. Children may have few or no symptoms and coarctation may go unnoticed for some time. Those children do not always need surgery, and if they do it is usually much less urgent than in infants.

The defect is rather easy to diagnose and the surgery is straight-forward. The surgeon places a clamp on either side of the narrowing, removes the narrowed section and restores the continuity of the aorta with sutures. This is not an open heart surgery because the surgeon is working outside the heart. The surgical results are excellent and more than 95% of patients will be able to lead a normal life after the operation.

In some cases, it is possible to enlarge the narrowed part by means of balloon dilation, thus avoiding any surgical procedure. Installing an endovascular stent may also help keep the vessel open.

After the operation, the hospital stay will be brief (approximately four to five days). In the case of balloon dilation, hospitalization often lasts less than two days. Children will be able to lead normal lives in most cases.

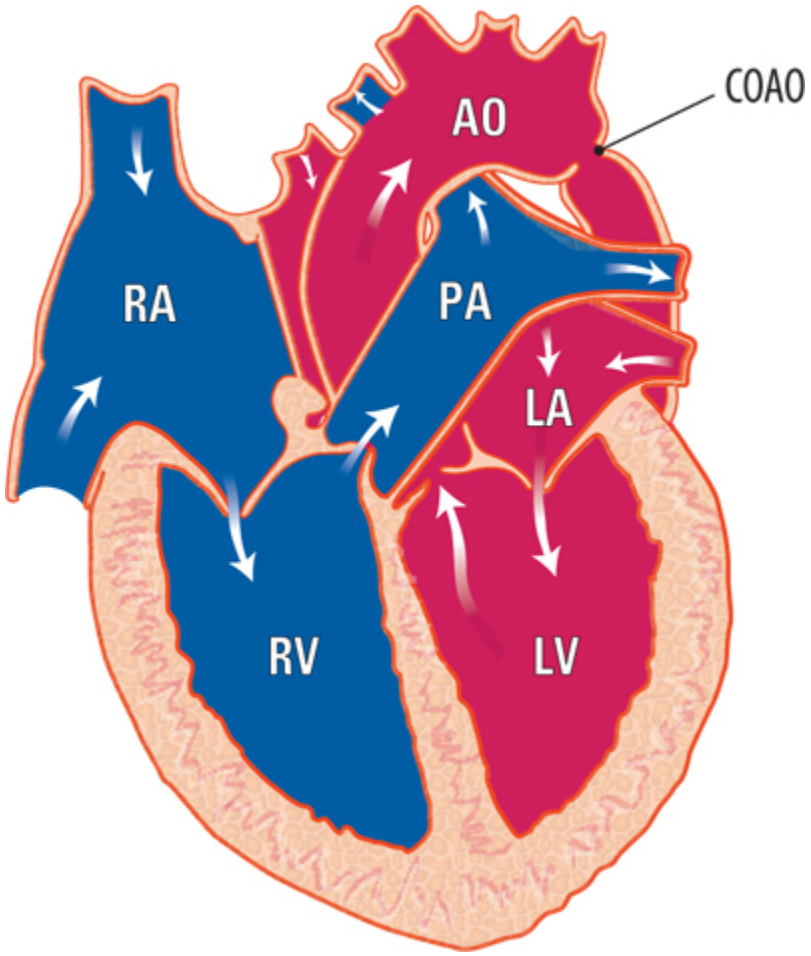


Figure 14 | Coarctation of the aorta

Atrioventricular septal defect

In the normal heart, a wall separates the two ventricles, and another wall separates the two atria. These two walls meet in the centre of the heart. This is also where the tricuspid valve ring and the mitral valve ring meet side by side. In atrioventricular septal defect (AVSD), also called complete atrioventricular canal, the location where the two walls merge has not developed normally. This causes an opening between the two ventricles as well as between the two atria. In addition, the development of the tricuspid and mitral valves is impaired, and a single large valve is usually found instead of two normal valves (Figure 15).

The consequences of this defect are the same as in the case of large ventricular septal defects (see the previous section on VSD). Thus, some of the oxygenated blood from the left side of the heart will travel back to the right side and will needlessly return to the lungs. Symptoms are also similar to those of ventricular septal defect (rapid breathing, difficulty with feedings and poor weight gain). In addition, because the tricuspid and mitral valves are not properly developed, they may leak. When the heart contracts, blood is thus returned to the atria. It is also possible for the mitral valve or the tricuspid valve to be too small or not open normally.

There are also “partial” forms of this defect in which only part of the wall between the atria is missing. Those partial forms are less severe and present as atrial septal defects (see section on atrial septal defect). They are sometimes accompanied by a mitral valve leak.

Except in some partial forms, atrioventricular septal defect requires cardiac surgery. When the defect is complete, the ideal time for corrective intervention is before the age of one year, usually around 6 months. When the defect only consists of an opening between the two atria (partial form), the operation will be done around 3 or 4 years old.

The operation consists in closing the openings between the atria and the ventricles by means of a synthetic patch and repairing the valvular defects to prevent future leakage. The results are usually good, and the children can live normal lives. However, repairing valves can be difficult and a leak may persist. Those residual leaks rarely require a new operation.

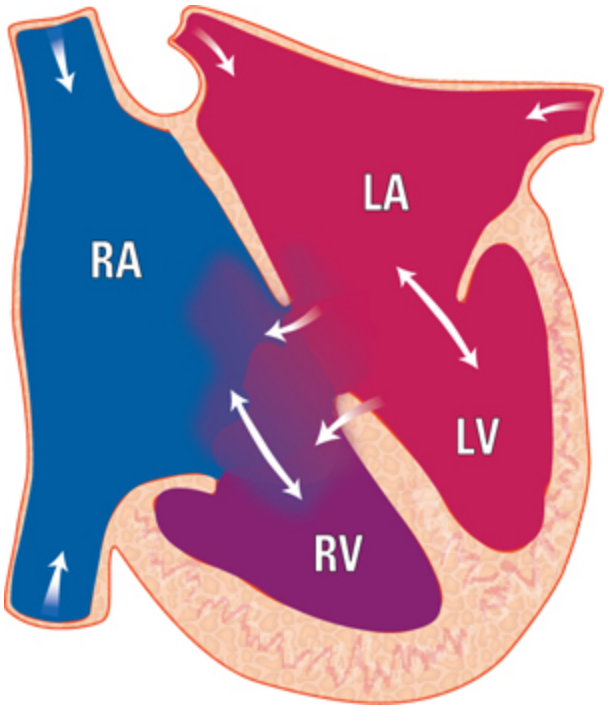


Figure 15 | Atrioventricular septal defect

Aortic valve stenosis

The aortic valve is located at the outlet of the left ventricle. It opens when the left ventricle contracts to allow blood to flow to the aorta. After the contraction, it closes to prevent blood from flowing back to the ventricle. Normally, when the heart contracts, the valves are large enough to let blood flow freely. In some circumstances, the valves developed abnormally and are too small. This is called stenosis. The normal aortic valve has three leaflets that open and close according to the heart contractions. In aortic stenosis, two of the three leaflets have often fused together, preventing them from doing their job properly. This is called a bicuspid aortic valve.

In aortic valve stenosis, the left ventricle must generate greater pressure to push blood through the malformed and undersized valve (Figure 16). Because the blood does not circulate well, the pressure is higher in the ventricle (before the valve) and lower in the aorta (after the valve). This pressure difference is called a “pressure gradient.” The tighter the valve, the higher the gradient. Pressures are measured in millimetres of mercury

(mmHg). This is the same unit of measurement as the blood pressure taken by the doctor. In the case of severe aortic stenosis, for example, the pressure may easily reach 220 mmHg in the left ventricle and 120 mmHg in the aorta. The gradient is then 100 mmHg (220-120).

In response to this extra work, the ventricle reacts like all muscles: it thickens. This is called hypertrophy. This hypertrophy allows the heart to continue to function despite the increased pressure, but this is not good in the long term.

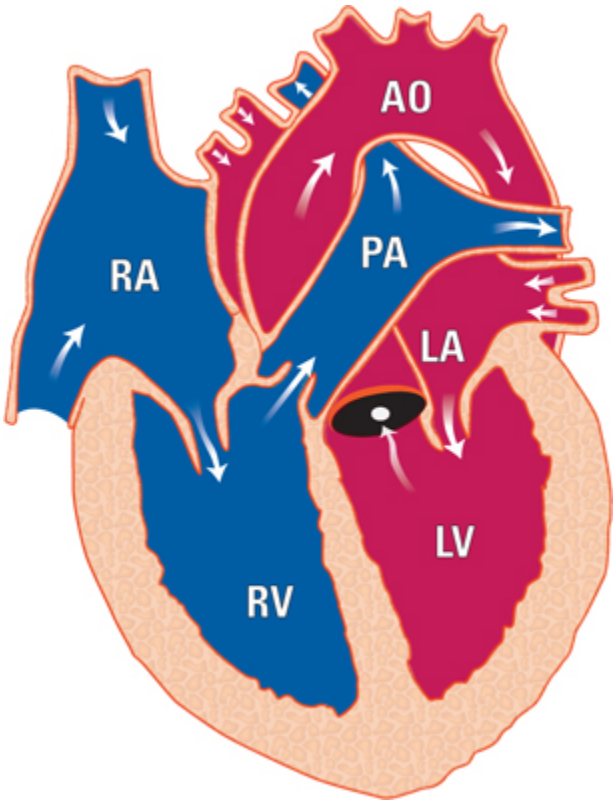
Fortunately, most cases of aortic stenosis are not very severe, they evolve slowly, and children do not exhibit symptoms. However, those children are regularly monitored to assess the progress of their stenosis. Some stenosis will remain mild for life, while others unfortunately get worse. If the stenosis becomes severe enough and the pressure difference is too great, or if the muscle becomes too thick, it is best to intervene.

In some cases, aortic valves that are too narrow can be dilated without surgery using a balloon-tipped catheter that is inserted into a large artery of the groin (the femoral artery). If dilation is not successful, or if the stenosis becomes severe again, open-heart surgery should be performed. In this case, the surgeon will try to repair the leaflets of the valve first. If that is not possible, he will have to replace the valve.

The malformed valve can be replaced with a mechanical valve. In some cases, the surgeon will opt for the Ross operation. This procedure involves replacing the child’s aortic valve with its own pulmonary valve (pulmonary autograft). The pulmonary valve is replaced by a human valve from a donor (homograft).

More rarely, the obstruction may be located directly below the valves (subvalvular aortic stenosis) or above (supravalvular aortic stenosis). Those types of stenosis may only be corrected surgically.

Figure 16 | Aortic stenosis



Pulmonary valve stenosis

The pulmonary valve is located at the exit of the right ventricle. It opens when the right ventricle contracts to allow blood to flow to the pulmonary artery. After the contraction, it closes to prevent blood from travelling back to the ventricle. When this valve is too narrow, the patient is said to have pulmonary valve stenosis (Figure 17).

In pulmonary stenosis, the right ventricle has to fight against the obstruction caused by the valve that is too narrow. In order to successfully push blood to the pulmonary artery and the lungs, the right ventricle must generate very high pressure as it contracts. In doing so, its wall thickens. Over time, the ventricle becomes unable to overcome the obstruction of the stenotic pulmonary valve and dilates. Its contraction becomes less and less effective. It is then said to fail.

This defect is often detected by the doctor who will hear a murmur in a child without symptoms. If the stenosis becomes severe and the ventricle has difficulty ejecting blood, exertion intolerance and shortness of breath may be observed. Sometimes pulmonary stenosis can be very severe from birth. The valve can also be completely closed (pulmonary atresia). The baby then has a bluish complexion (cyanosis) and may develop severe symptoms soon after birth.

When the opening is too narrow, it must be enlarged. Usually, this can be done without surgery using a balloon catheter. This catheter is inserted through a vein in the groin. The balloon is briefly inflated at the level of the malformed valve in order to dilate it and thus restore normal circulation. This intervention can be done at any age, even when the stenosis is very narrow, including during the neonatal period. However, in some cases the valve becomes stenotic again and further intervention is needed. The transcatheter dilation may be performed again, but sometimes heart surgery is required.

When the pressure in the right ventricle is not high enough to justify a procedure, the child simply has to be monitored periodically with an echocardiogram to make sure that the stenosis is not worsening with time. Patients living with pulmonary stenosis tend to have normal lives and the results of procedures, when needed, are generally excellent.

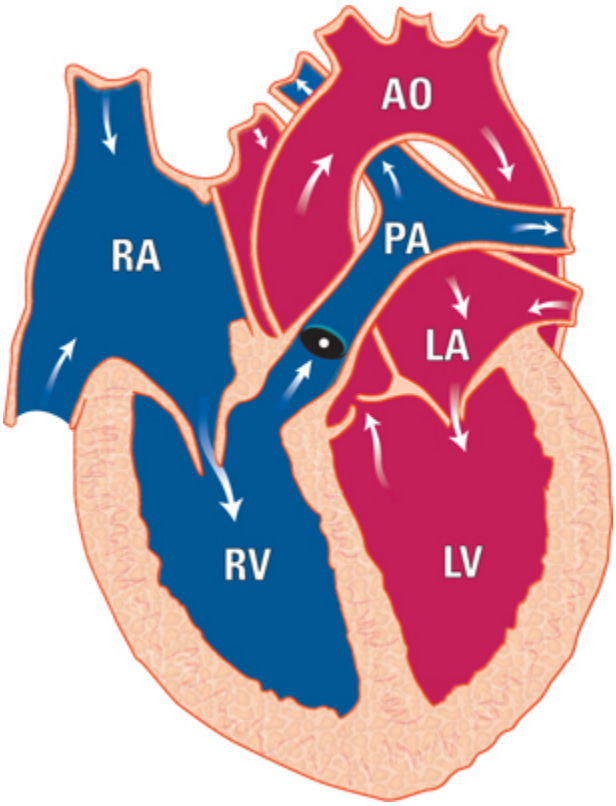


Figure 17 | Pulmonary stenosis

Tetralogy of Fallot

Tetralogy of Fallot is a fairly complex defect. In Étienne-Louis Arthur Fallot's original description of it in 1888, it included four defects: a large opening between the two ventricles (ventricular septal defect), a narrowing between the right ventricle and the pulmonary artery (pulmonary stenosis), a forward shift of the aorta and, ultimately, a thickening of the wall of the right ventricle (ventricular hypertrophy). The tetralogy of Fallot does not comprise four different defects. Rather, it is a single anomaly in the development of the heart that causes these four defects. Early in the development of the heart, the wall that separates the aorta from the pulmonary artery develops a little too close to the front of the heart. This pulls the aorta forward, thereby obstructing the outlet of the right ventricle. This shift is also responsible for the misalignment of the walls of the heart and causes a wide opening between the two ventricles. The last defect

described by Fallot, the right ventricular hypertrophy, is not always present at birth, but develops over time in children without surgery (Figure 18).

Tetralogy of Fallot is a common heart defect (8% of all heart defects). This is the most common cyanogenic defect, which means that blue (deoxygenated) and red (oxygenated) blood are mixed in the circulation. This is because the blue blood from the right ventricle cannot easily travel to the lungs due to pulmonary obstruction (stenosis). Since there is a large opening between the ventricles, the blue blood travels through the aorta to mix with the red blood ejected from the left ventricle. This mixture of blue and red blood may give the child a bluish complexion (cyanosis). The cyanosis will be more or less pronounced depending on the severity of the obstruction between the right ventricle and the pulmonary artery.

This defect often goes unnoticed at birth because pulmonary stenosis develops progressively. Children gradually turn blue after a few days or weeks. Today, however, the most tetralogies of Fallot are detected either during pregnancy or during examination of the newborn.

This defect must be corrected with open-heart surgery. During the operation, the surgeon closes the ventricular septal defect with a synthetic patch and then opens the regions of the right ventricle and of the pulmonary artery that are obstructed. The operation is usually performed around 6 months (sometimes earlier in some more severe cases). In general, children who are operated go on to have normal lives. However, sometimes the pulmonary valve is so obstructed that the surgeon has to remove it completely, causing the valve to leak. This leakage is well tolerated in childhood, but over time it can cause a dilation of the right ventricle. This dilation can lead to exertion intolerance and sometimes severe arrhythmia. It may therefore be necessary to replace the pulmonary valve during adolescence.

In an unoperated child, the part of the right ventricle leading to the lungs may go into spasm when experiencing strong emotion or anger. This forces almost all the blood to pass from the right ventricle to the aorta without flowing through the lungs. The child suddenly turns very blue. This is called a cyanotic or blue spell (hypoxia). The cyanotic spell often resolves

spontaneously, but it may also be serious if it persists. In the presence of cyanotic spells, surgery will be performed earlier since repairing the defect resolves the problem.

In certain severe cases, the pulmonary arteries may be absent or quite underdeveloped (tetralogy with pulmonary atresia). The child then needs an early first surgery. The repair is mostly done in stages. An attempt is made to connect the segments of the pulmonary arteries of sufficient diameter to each other in order to rebuild a functional pulmonary arterial network which is then connected to the right ventricle using a prosthesis (homograft or conduit). In those cases, complications are unfortunately more frequent.

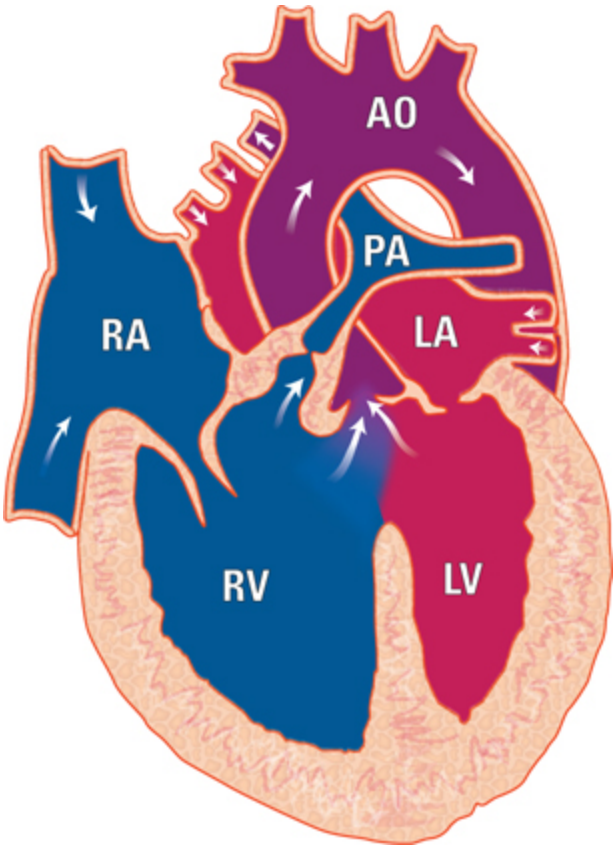


Figure 18 | Tetralogy of Fallot

Double outlet right ventricle

Double outlet right ventricle occurs when the two main arteries of the heart, the aorta and the pulmonary artery, arise from the right ventricle. It is a complex defect of which there are several variants. When a pulmonary obstruction occurs, it is said to be a double outlet right ventricle of the tetralogy of Fallot type. Symptoms and treatment are similar to the tetralogy of Fallot (see previous section). A double outlet right ventricle without pulmonary valve obstruction causes symptoms similar to those of large ventricular septal defect. All forms of double outlet right ventricle have a ventricular septal defect that may be located at different sites along the interventricular wall.

The surgery consists essentially of redirecting the blood from the left ventricle to the aorta, closing the ventricular septal defect and relieving the pulmonary stenosis, if present. This surgery can be done early and usually gives good results. There are also other, rarer forms with defects in the positioning of the great vessels and which may be more difficult to repair.

Transposition of the great vessels

Transposition of the great vessels is a defect in which the two arteries arising from the heart are inverted. Thus, the aorta originates from the right ventricle and the pulmonary artery from the left ventricle (Figure 19). If we follow the blood flow, we realize that the blood returning from the lungs (oxygenated) drains into the left heart, but then returns to the lungs without passing through the rest of the body. Likewise, blue (deoxygenated) blood from the organs enters the right atrium and then the right ventricle, exits the aorta and returns to the organs without passing through the lungs. The blood in the aorta is blue, causing cyanosis in the newborn (bluish-coloured baby). Therefore, there is not enough oxygen going to the organs.

When the two circulations are completely independent, life is not possible. Survival therefore depends on mixing the two circulations. Mixing may be achieved when blood flows through the ductus arteriosus, atrial septal defect or ventricular septal defect. Often, emergency medical interventions are essential to save the infant’s life by allowing an exchange of blood

between the two circulations. Administration of medication intravenously will keep the ductus arteriosus open. In addition, an urgent opening of the atrial septum through cardiac catheterization is usually necessary. Some blue blood will therefore travel to the lungs and some red blood will travel to the organs.

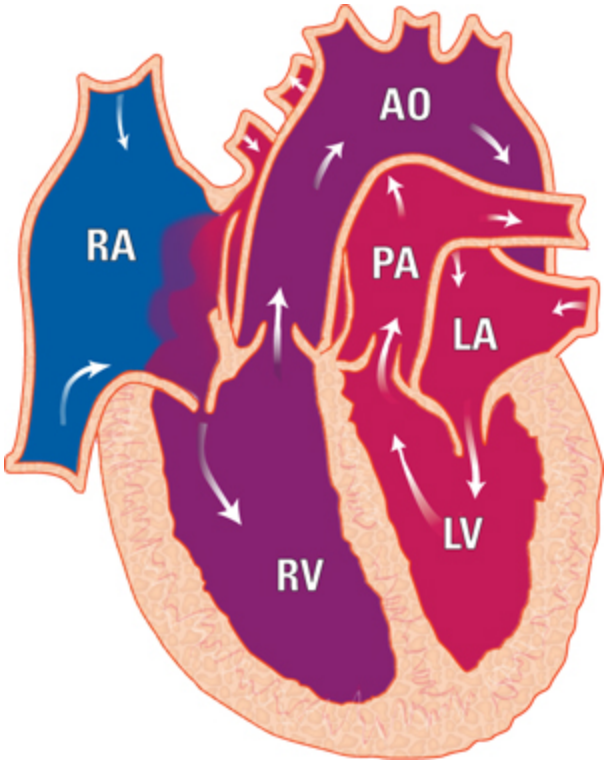


Figure 19 | Transposition of the great vessels

An infant with this defect will need heart surgery in the first few days of life. The surgery first involves cutting the aorta just above the aortic valve and the main pulmonary artery just above the pulmonary valve, then switching these two arteries to connect them in the right place. The coronary arteries are also moved to follow the aorta. Openings such as ductus arteriosus and atrial septal defect are then closed. The surgery is delicate, but usually goes well. Children who are operated lead normal lives afterwards and few have long-term complications.

Truncus arteriosus

Truncus arteriosus is a rare cyanogenic defect (blue baby syndrome). It is characterized by only one artery arising from the heart instead of two, hence the name common arterial trunk or truncus arteriosus (Figure 20). This common arterial trunk divides to create the pulmonary arteries and the aorta. In addition, there is always a large ventricular septal defect. Blue (deoxygenated) blood from the right ventricle and red (oxygenated) blood from the left ventricle mix in the common arterial trunk. As a result, there is some blue blood that ends up in the aorta. Since the pressure in the pulmonary arteries is lower than in the aorta, the blood leaving the heart tends to go to the lungs. This overflow of blood to the lungs causes rapid breathing.

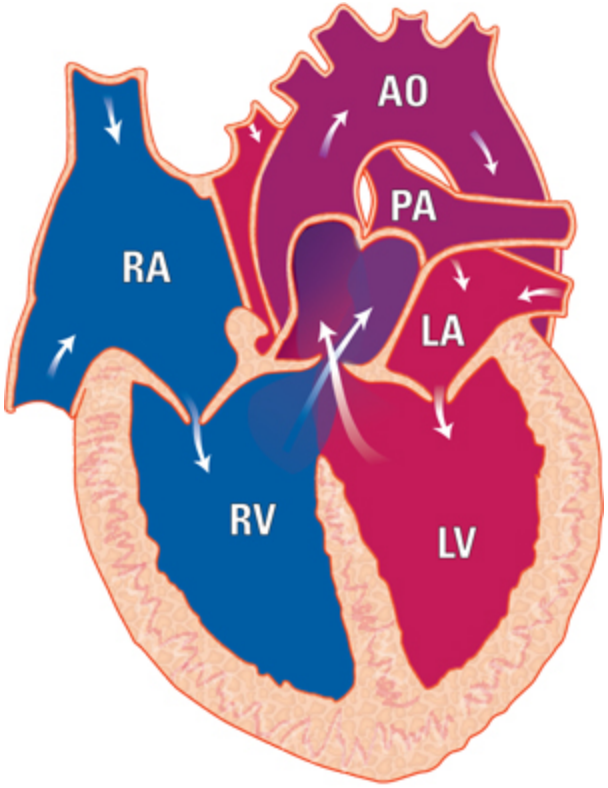


Figure 20 | Truncus arteriosus

The physical signs (cyanosis and rapid breathing) of this defect appear within the first days of life. To avoid complications and to improve the condition, the newborn baby should undergo early heart surgery. This involves closing the opening between the ventricles, separating the pulmonary arteries from the common arterial trunk and placing a conduit between the right ventricle and the pulmonary arteries. Later in life, the

conduit must be replaced, as it becomes too small for the growing child. Usually, the operated child is able to do the same activities as other children. In some cases, the trunk valve leaks, or its opening is too narrow and it also needs to be corrected.

Single ventricle or univentricular heart

Single ventricle refers to a set of serious heart defects that have in common the fact that only one ventricle is functional instead of two. This happens when one of the ventricles has not developed properly, or when the mitral or tricuspid valve is missing or too small. In reality, there are often two ventricles, but only one is truly functional, hence the name univentricular heart. An example of a univentricular heart is shown in Figure 21. Hypoplastic left heart and tricuspid atresia are examples of univentricular hearts (see below).

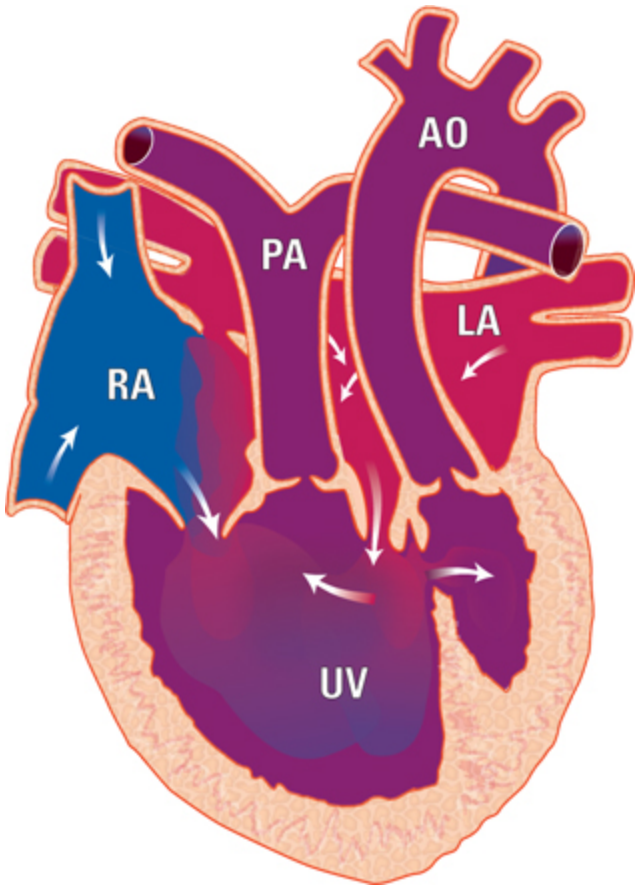


Figure 21 | Example of univentricular heart

Children with such defects are almost always cyanotic and may also have heart failure. They will need surgery more than once. Since it is nearly impossible to reconstruct both ventricles, at least two procedures will be required to allow blood to flow passively to the lungs, without passing through the heart. The blood then returns from the lungs to the heart and it is pumped by the only functioning ventricle to the organs of the body. The first operation, called a superior cavopulmonary anastomosis (or Glenn procedure), is performed before the age of one. The second operation, called Fontan operation, is performed between the ages of 3 and 5. In some cases, surgery soon after birth is also necessary while waiting for the Glenn procedure to ensure that the blood supply to both lungs and to the other organs of the body is adequate. Some children can live almost normally with a univentricular heart, but the risks of complications (exertion intolerance, cyanosis, arrhythmia and heart failure) remain high. However, the care of children with univentricular heart is changing rapidly and many adults now live with this type of defect.

Hypoplastic left heart syndrome

In hypoplastic left heart syndrome, the left side of the heart is underdeveloped. The left ventricle is tiny or nonexistent, the aortic valve is often imperforate, the left atrium is small, and the aorta is underdeveloped. Therefore, all the blood coming from the lungs throughout the pulmonary veins cannot go to the left ventricle: it must flow through the right atrium through an opening in the interatrial septum. It then travels to the right ventricle, returns to the pulmonary artery and through the ductus arteriosus to the aorta to supply blood flow to the rest of the body (Figure 22). Because the ductus arteriosus naturally closes in the first few days of life, the child can quickly become very ill and even die.

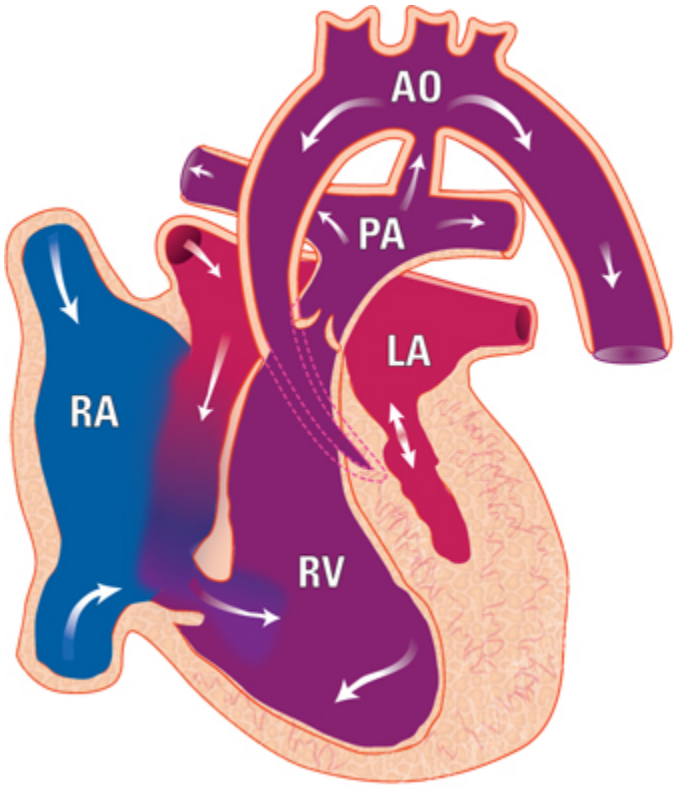


Figure 22 | Hypoplastic left heart

The symptoms appear within the first hours or the first few days of life. The newborn experiences shortness of breath, cyanosis, is pale and sometimes lethargic. Emergency treatment involves giving the newborn a medicine called prostaglandin which will keep the ductus arteriosus open, enabling the baby to survive until the first operation. This complex procedure, called the Norwood operation, involves fusing the main pulmonary artery (normal size) with the aorta (too small) to create a new aorta of appropriate size. The arteries that travel to the lungs are then detached from the main pulmonary artery and a small conduit is placed between them and a branch of the aorta to deliver blood to the lungs.

Two additional surgical procedures will be necessary before the age of 5 to complete the reconstruction of the univentricular heart (see the previous section on univentricular hearts). Overall, the risks of mortality and complications resulting from this defect are unfortunately high.

Tricuspid atresia

This is a relatively uncommon cyanotic heart defect (bluish-coloured child) in which the tricuspid valve is nonexistent (Figure 23). The blood that comes from the inferior and superior vena cava flows into the right atrium and, unable to reach the right ventricle, goes across an opening in the interatrial septum into the left atrium and the left ventricle. The latter returns blood to the aorta, but also to the small right ventricle through a ventricular septal defect. The symptoms of this defect depend on the development of the pulmonary valve and pulmonary arteries. If they are too underdeveloped or stenotic, the newborn may quickly become cyanotic and should be given medication to keep the ductus arteriosus open. In other cases, the circulation of blood to the lung is not obstructed and will even become too abundant, which will cause the child to experience shortness of breath.

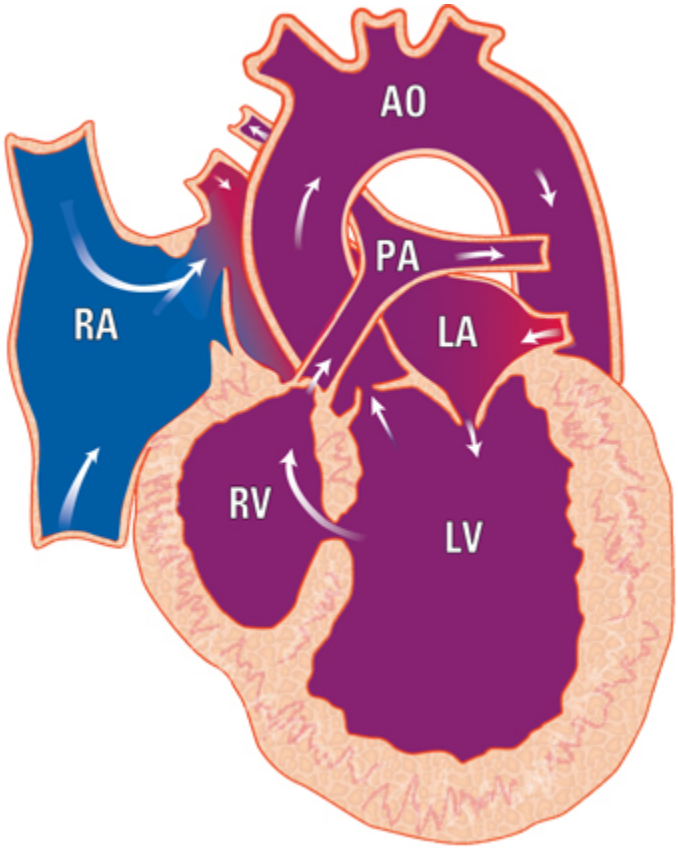


Figure 23 | Tricuspid atresia

On the one hand, if there is not enough blood flowing to the lungs, an initial surgery soon after birth will be essential to place a synthetic tube between a branch of the aorta and the pulmonary artery. On the other hand, if there is too much blood flowing to the lungs, it will be necessary to install a band to constrict the pulmonary artery and limit the amount of blood that passes through it.

Since the right ventricle will never be functional, two more operations will be necessary in order to rebuild a univentricular type heart (see the previous section on univentricular hearts).

Total anomalous pulmonary venous return

In a normal heart, the oxygenated blood that exits the lungs returns to the left side of the heart through four pulmonary veins (two on the left and two on the right). These veins are normally connected to the left atrium. In the total anomalous pulmonary venous return defect, all four veins drain abnormally into a common collecting vessel that eventually reaches the right atrium (Figure 24) instead of connecting to the left atrium. The oxygenated blood returning from the lungs is therefore sent to the right side of the circulation. The connection may be made directly to the right atrium. In some other cases, the collecting vessel descends below the diaphragm, enters the abdomen and then ascends through the liver. The amount of blood returning to the right side of the heart is therefore greatly increased and much of the oxygenated blood returns to the lungs to be needlessly oxygenated again.

This defect results in oxygenated red blood mixing with blue blood in the right atrium. Some of this mixture returns to the left atrium through an opening between the two atria and then travels to the aorta. The child therefore has a decreased oxygen saturation and is thus cyanotic.

When the diameter of this collecting conduit is large enough, the child may survive for months without too many problems. However, he may experience shortness of breath and not grow normally. Unfortunately, in

many cases, this collecting conduit is narrowed at the end. This is called anomalous pulmonary venous return with obstruction: the blood can no longer exit the lungs. In this case, the newborn becomes ill within the first days of life and requires urgent surgery.

This defect is corrected surgically as soon as it is detected. The operation consists in reconnecting the pulmonary veins to the left atrium to restore normal circulation. The results are generally excellent, and the child can expect to have a normal life without any impairment. Exceptionally, excess scarring may occur and the area where the pulmonary veins have been reconnected may narrow. It is therefore necessary to correct this area again, sometimes with a balloon catheter or a second surgery.

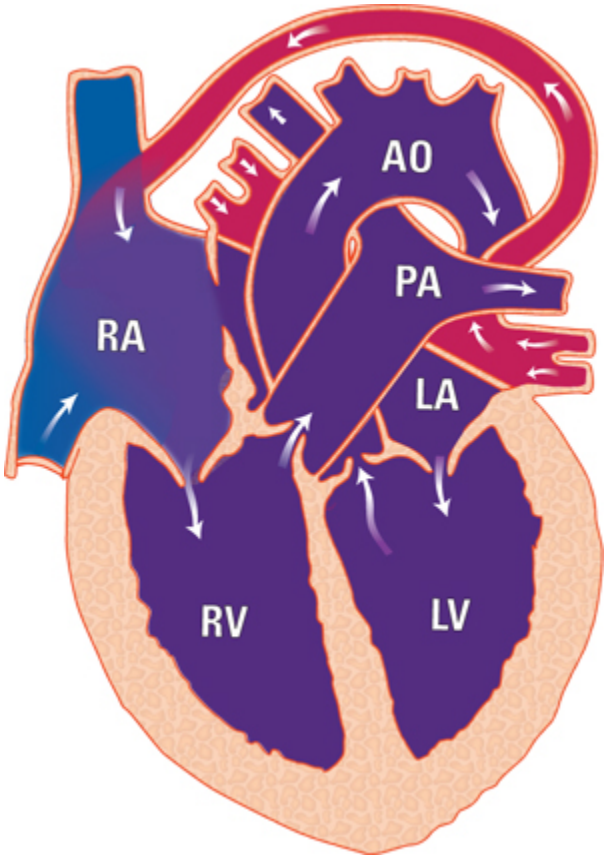


Figure 24 | Total anomalous pulmonary venous return

Partial anomalous pulmonary venous return

This defect causes some pulmonary veins to abnormally drain to the right atrium instead of the left atrium. Oxygenated blood from these veins returns to the right heart either through an abnormal connection directly to the right atrium or through a vein already receiving deoxygenated blood (Figure 25). Depending on the number of abnormal veins, the defect has more or less important consequences. Circulation occurs in a manner similar to an atrial septal defect. There is consequently a lot more blood returning to the right ventricle and to the lungs. This defect may go unnoticed.

If there is only one abnormally connected pulmonary vein, the defect may not cause symptoms. It is not always necessary to correct it. If there are symptoms, or if two or more pulmonary veins are abnormal, open-heart surgery is recommended. Surgery is usually offered before school age or earlier if symptoms develop (shortness of breath following exertion, difficulty gaining weight, or frequent respiratory infections). Often times, this defect occurs with another heart defect, such as atrial septal defect. When they are present together, the two defects are corrected at the same time. The results of the surgery are excellent, and the child can expect to live a completely normal life.

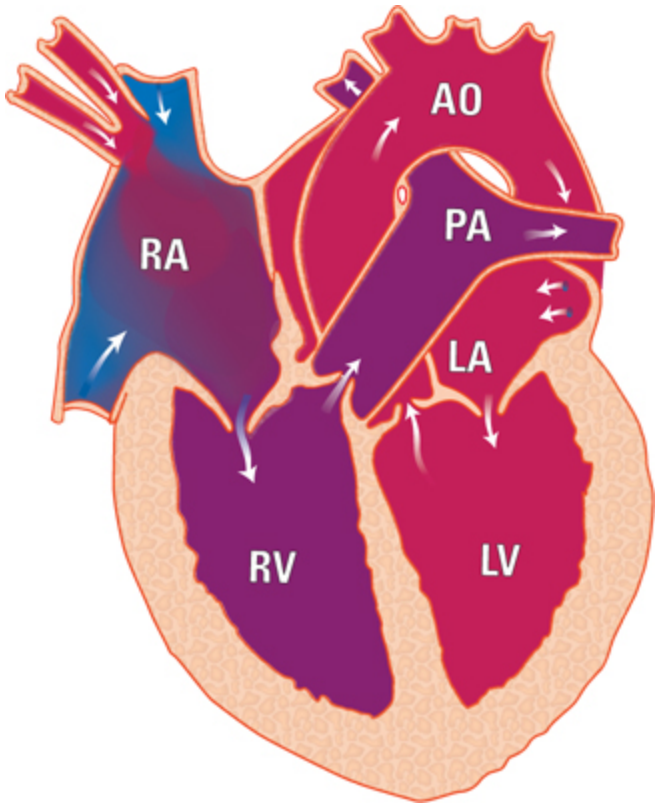


Figure 25 | Partial anomalous pulmonary venous return

Ebstein’s anomaly

Ebstein’s anomaly is characterized by a defect of the tricuspid valve (the valve inside the right heart between the atrium and the ventricle). Because its leaflets adhere to the walls of the right ventricle and its opening is displaced down the ventricle, the valve may not close normally (Figure 26). The severity of the defect varies, and therefore symptoms and treatments vary accordingly. In the very severe form of the disease, the tricuspid valve is so displaced inside the right ventricle that there is hardly any right ventricle left to ensure that blood is pumped to the lungs. When the tricuspid valve fails to close properly, blood collects in the right atrium as the right ventricle contracts (blood that would normally go into the lungs returns to the atrium). The right atrium then gradually enlarges and arrhythmia (disturbances in the electrical system of the heart such as tachycardia) may occur.

In addition to their tricuspid valve defect, it is also common for children with Ebstein's anomaly to have an additional electrical pathway that may lead to tachycardia. It is seldom necessary to intervene at a very young age, but, in general, these children will need to have their tricuspid valve replaced or corrected. It is a complex surgery, and it is not always possible to correct the defect completely. The child's abilities and complications will vary depending on the severity of their defect and the results of their surgery.

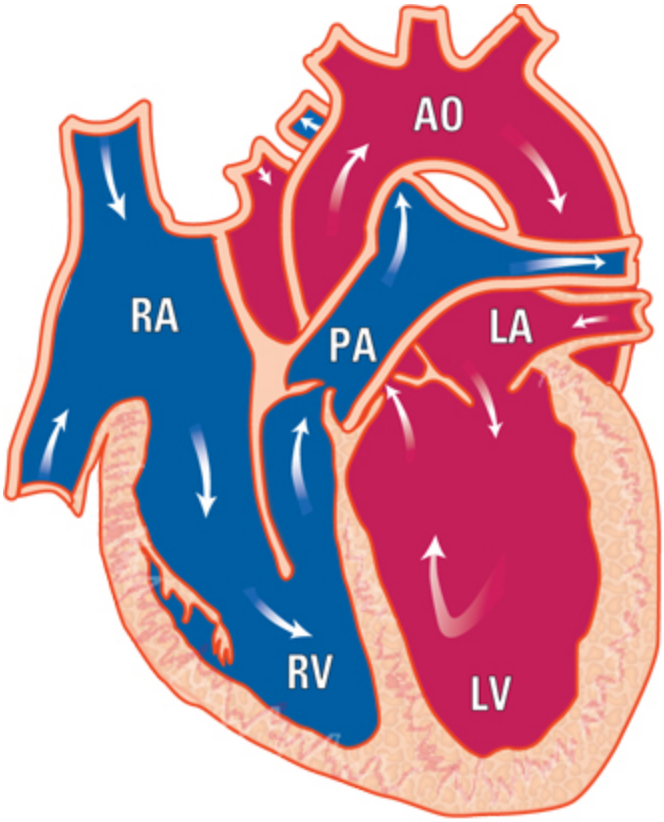


Figure 26 | Ebstein's anomaly

Vascular ring

A vascular ring occurs when the main arteries of the heart (the aorta and its branches) do not develop normally and form a ring that encircles the trachea (the conduit that brings air to the lungs) and the esophagus (the tube which brings food to the stomach). There are different types of vascular

rings. The two most common are the double aortic arch and the right aortic arch with aberrant left subclavian artery.

The normal aorta arches and then plunges to the left. This is called left aortic arch. The first vessels that arise from the aorta are, in order, the brachiocephalic artery, the left carotid artery and the left subclavian artery.

In the right aortic arch, the aorta rather plunges to the right and the vessels emerging from it mirror those of a normal aorta. This usually does not cause any problem, but there may be a left subclavian artery that arises abnormally right after the right subclavian artery. This left subclavian artery travels behind the trachea and esophagus, trapping them behind the aorta.

In the double aortic arch, the aorta divides into two as it exits the left ventricle. These two aorta segments encircle the esophagus and trachea and then merge again to form the descending aorta. The esophagus and the trachea are thus caught between the two arches (Figure 27).

In other types of vascular rings, the anatomy of the great vessels varies, but the principle remains the same: the esophagus and trachea are squeezed between two structures and cannot develop normally. In the majority of cases, vascular rings are not a problem and are often diagnosed by chance. However, when the ring causes an obstruction of the trachea or of the esophagus, the child may exhibit difficult and noisy breathing, have trouble eating or swallowing, have pain when swallowing, feel that something is stuck in his throat, feel like he is choking, or suffer from a chronic cough or acid reflux.

Several tests may be necessary to properly diagnose a vascular ring: an X-ray of the lungs, an echocardiogram, a bronchoscopy or a cardiac magnetic resonance. Usually, no treatment is required if there are no symptoms. If the vascular ring is causing significant symptoms, the child will need to have surgery to correct or remove the obstruction. In most cases, surgery resolves the problem, and the child can then live a normal life.

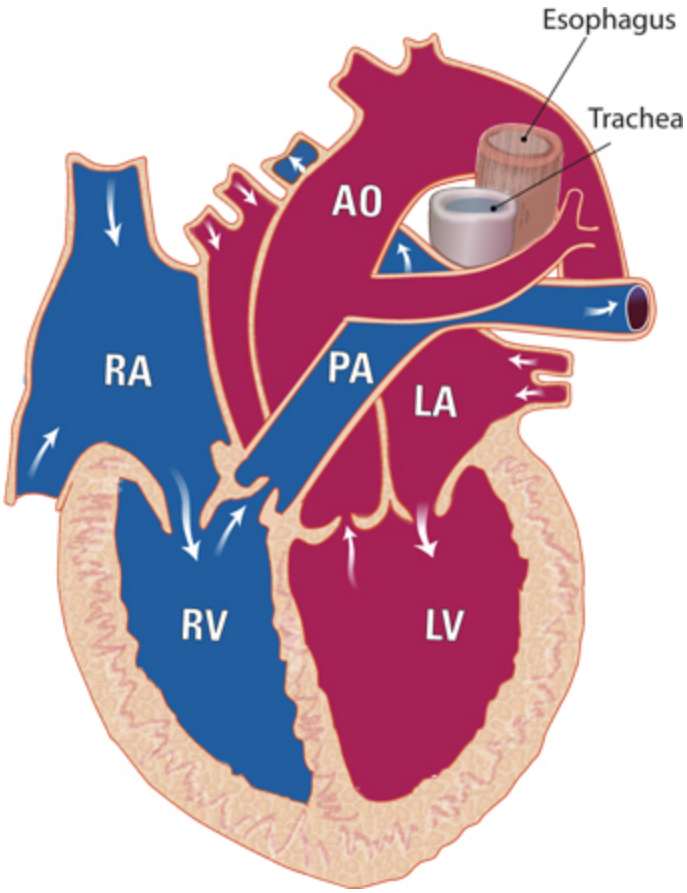


Figure 27 | Vascular ring

Cardiomyopathy

Cardiomyopathy is a disease affecting the muscle of the heart called the myocardium. In cardiomyopathy, the heart muscle becomes too thick, too stiff or too thin, which affects its ability to properly fill and pump blood.

There are several causes of cardiomyopathy and they are sometimes difficult to identify. Myocarditis (an infection that affects the muscle of the heart), chemotherapy, a metabolic or muscle problem, or genetic problems are the most common causes in children. In some cases, cardiomyopathy is caused by diabetes or a myocardial infarction. However, these causes are very rare in children and are usually found in adults. There are different types of cardiomyopathy.

Dilated cardiomyopathy

It is the most common type of the disease, in both children and adults. The chambers of the heart get too large because the weakened heart muscle has difficulty pumping blood. Some children may have no symptoms while others may have heart failure. In infants, heart failure results in difficult breathing, difficult feeding, excessive sweating and difficulty gaining weight. There are several causes that may lead to dilated cardiomyopathy and some may be hereditary, i.e., transmitted through family genes.

Hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy occurs when the heart muscle becomes abnormally thick. The thickening of the walls prevents the ventricles from filling properly and hinders the flow of electricity through the heart which may cause severe arrhythmia. This type of cardiomyopathy is often inherited.

Restrictive cardiomyopathy

This type of disease is rare in children. The walls of the heart become too rigid, which prevents the organ from relaxing and filling. In more severe cases, the drugs are often ineffective, and the only treatment is heart transplantation.

Ventricular non-compaction or spongiform cardiomyopathy

This form of the disease is also rather rare. It consists of a defect in the development of the heart muscle. Instead of becoming solid and strong, it remains porous as it was in the fetus (hence the term “spongiform,” which suggests a sponge-like texture). Heart muscle that has not compacted gradually expands and may eventually lose its ability to pump blood, as in dilated cardiomyopathy described above. In other cases, the heart becomes exaggeratedly thick as in the hypertrophic form. The causes for this defect

are still poorly understood. Symptoms vary widely. Some children will have no symptoms, while others will be severely affected and will need a heart transplant.

Treating cardiomyopathy is not always straightforward. Depending on the form of cardiomyopathy, specific medication may reduce cardiac dilation, help the heart relax, fill and pump blood. It may also decrease arrhythmia. Often a combination of drugs must be used.

In more severe cases, the insertion of a defibrillator will be necessary. A defibrillator is a small device placed under the skin which is connected to the heart by electrodes. It controls the rhythm of the heart to prevent severe arrhythmia. Sometimes, the surgeon removes excess muscle that prevents the heart chambers from filling properly. Unfortunately, transplantation is sometimes essential when heart failure progresses or does not respond to treatment.

Arrhythmia

As we saw earlier in the section “The control of the cardiac system,” the heart’s electrical system allows it to beat regularly and in an orderly fashion by passing an electrical impulse from the atria to the ventricles. This impulse tells the heart muscle when to contract and when to relax. The pulse usually begins in the sinus node, which is located in the right atrium. The sinus node is the heart’s natural pacemaker (Figure 28). The current first travels through both atria before reaching the central part of the heart, between the atria and the ventricles. This central part is called the atrioventricular node, or AV node. This is normally the only electrical connection between the atria and the ventricles. After passing the AV node, the electrical impulse divides into two branches to reach the right and left ventricles and thus produce their muscular contraction.

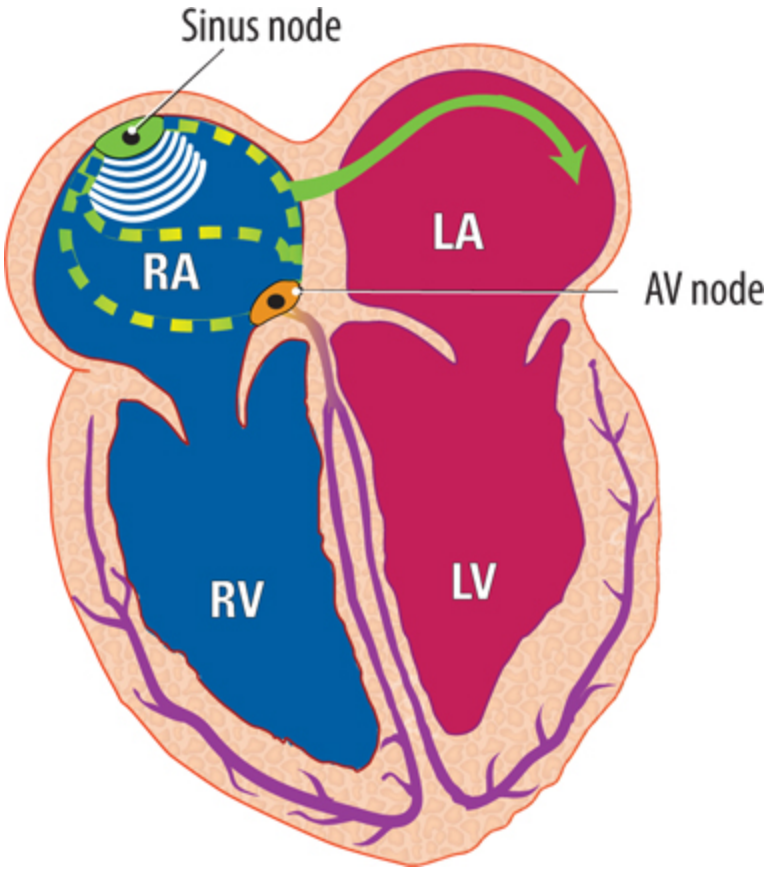


Figure 28 | Electrical system of the heart

Some problems may occur in the heart's electrical system, leading to specific consequences, including arrhythmia. Those are detailed in the following pages.

Atrioventricular block

Atrioventricular block, or AV block, occurs when electricity does not travel well through the AV node. The electrical impulse therefore does not pass correctly from the atria to the ventricles, as conduction may be slowed down or blocked completely.

There are several types of AV blocks. It may be congenital, i.e., present in the fetus during pregnancy or at birth. It is then associated with heart defects or the presence of autoimmune diseases in the mother such as lupus erythematosus. AV block may also be acquired, meaning it appears after birth and may be the result of different causes (a scar, damage to heart

tissue, surgery, certain medications, or inflammation of the heart caused by a virus or disease).

There are three degrees of AV block. In the first degree, electricity flows more slowly than usual through the AV node, but still travels to the ventricles. The second degree takes two different forms. In the first form, the time it takes for the electrical impulse to pass through the AV node gradually increases over a few beats until one of them is completely blocked (not passing from the atria to the ventricles). In the second form, some of the electrical impulses are completely blocked on a regular basis. Finally, in the third degree, also called the complete AV block, electricity does not pass through the AV node at all. There is therefore no more electrical communication between the atria and the ventricles.

The majority of patients have no symptoms. Patients most likely to exhibit symptoms are those with complete AV block, especially if the beating rate of the ventricles is slow. Symptoms may include syncope (loss of consciousness), dizziness, lightheadedness, extreme fatigue, shortness of breath, palpitations or a feeling of missing heartbeat, or intolerance to exercise.

AV block is usually diagnosed with an electrocardiogram or a Holter (24-hour electrocardiogram recording). First degree AV block and some second-degree blocks are mild and do not need treatment. However, patients with complete AV block will need a cardiac pacemaker to make sure their hearts are beating fast enough. The pacemaker is a small device located under the skin which is connected to the heart by wires. It is through these wires that electrical waves are sent to the heart, giving an electrical impulse to the atria and ventricles. This re-establishes and synchronizes the basic rhythm between the atria and the ventricles.

Supraventricular tachycardia

Supraventricular tachycardia (SVT) is a problem with the rhythm of the heart. The term “tachycardia” means “fast heart.” In the majority of cases, the increased heart rate that defines SVT is due to abnormal electrical

communication between the atria and ventricles or even within the AV node itself. It is the most common arrhythmia in children.

In SVT, the heartbeat may be so fast that the heart becomes less efficient at pumping blood. In this case, various symptoms may occur: heart palpitations (uncomfortable feeling of fast beating or sudden changes in heart rate), chest pain, dizziness or even syncope (loss of consciousness). Infants may also have difficulty breathing, feeding, irritability and vomiting.

Diagnosis is often made using one of the tests that record the heart's electricity, usually an electrocardiogram (recording for a few seconds) or a Holter (24-hour ECG recording). Home ECG transmitters are also regularly used. This is a device that you keep on your person and stick to your chest when you experience heart palpitations. The device then records the heart's electricity during the palpitations, which helps the cardiologist make a diagnosis.

Most SVTs that occur before or soon after birth tend to go away with time. Treatment may then be stopped. However, some children may continue to have episodes of SVT and will require long-term treatment.

There are three types of treatment: vagal maneuvers, medication, and ablation. Vagal maneuvers may be attempted to stop an episode of SVT. They include: holding your breath, "pushing" to inflate the stomach, blowing through a blocked straw, applying ice to the face, coughing or vomiting. Various medications may also be used to decrease episodes. The choice of medication will be based on the child's age and type of arrhythmia. Finally, in older children, a procedure to "burn" the small electrical short circuit that causes tachycardia episodes may be performed. This is called an ablation. Using tiny catheters inserted into the veins of the groin, the electrophysiologist (a cardiologist who specializes in arrhythmia) burns the abnormal electrical cells. Most children can go home the day after the procedure.

Medical follow-up depends on the patient's age and the type of episode. Babies will need to be followed more closely during the first year of life but

may not need to be followed afterwards if SVT goes away completely. Children in whom episodes are not very well controlled will require more frequent monitoring. Children with a successful ablation may not need further follow-up. The majority of children with SVT lead normal, unrestricted lives.

Kawasaki disease

Kawasaki disease is characterized by a high fever with inflammation (redness or swelling) of the hands, feet, mouth, eyes and other parts of the body. The blood vessels may also be affected by inflammation. The coronary arteries are most often affected. These are small vessels used to deliver blood and oxygen to the muscle of the heart. If this inflammation is treated quickly, children recover quickly and usually have very little further damage. However, if the disease is severe or is not treated promptly, it may result in damage to the heart, mainly by causing aneurysms (widening) of the coronary arteries.

No one really knows what exactly causes Kawasaki disease, but many believe it is an abnormal reaction of the immune system to a virus or bacteria. Kawasaki disease is not contagious. People of Asian descent are more at risk of getting the disease. In fact, the name “Kawasaki” comes from the Japanese doctor who first described this disease.

There are no specific tests to diagnose Kawasaki disease. Rather, the diagnosis is based on signs and symptoms as well as certain lab tests showing signs of inflammation. When this disease is suspected, an assessment is usually performed by a cardiologist, who conducts an electrocardiogram and an echocardiogram.

The child must be admitted to the hospital to receive immunoglobulins, a medicine that can only be given intravenously. Immunoglobulins are proteins from blood products that act as antibodies to decrease the body’s abnormal immune response. In some cases, immunoglobulins are not sufficient and other drugs such as steroids and immunosuppressants must be

used. When the fever subsides and the child’s condition improves, an observation period of at least 24 hours is usually necessary to ensure that the fever and inflammatory response do not return.

In the majority of cases, there is no serious damage to the coronary arteries and the medication is quickly discontinued. If aneurysms or dilation of the coronary arteries develop, aspirin or other medicines to prevent the blood from clotting may be prescribed for a longer period. Severe damage is rare. If this is the case, the child should be monitored regularly. It is then possible that interventions or surgery are required.

Chapter 5

The hospital and treatments

The monitoring of children with a heart defect is usually handled in an outpatient clinic. Depending on the type of defect, the frequency of appointments may vary. There may also be a need for hospitalization, diagnostic tests, catheterization procedures and surgery. It is therefore useful and reassuring to become familiar with the way hospitals operate, both in the outpatient clinic and in the inpatient departments, in the operating room and the intensive care units. A good knowledge of the role of each of the principal caregivers is also essential.

The outpatient clinic

The majority of appointments will be handled in an outpatient clinic. Most pediatric cardiology centres try to organize their clinic by performing all the necessary tests in a single visit. Therefore, an outpatient visit may last a few hours. Common outpatient diagnostic tests include electrocardiograms, echocardiograms, stress tests and blood tests. In most outpatient clinics and when possible, the patient always sees the same cardiologist. The frequency of outpatient visits depends a lot on the child’s age and the type of heart disease. A newborn will be seen more frequently, for instance, than an older child whose visits may be several years apart. It should be added that pediatric cardiologists often provide an outreach clinic service in regional hospitals close to home. One or two cardiologists will travel to the region, sometimes accompanied by an echocardiography technician. The list of these outreach clinics is available in the various university hospital centres.

It is best to check with the cardiologist to see if such follow-up may be scheduled in one of these clinics. It is important to note that the En Cœur Foundation has been supporting these outreach clinics for many years, allowing their deployment in a growing number of regional hospitals across Quebec.

Hospitalization

The child with a heart problem will rarely be hospitalized. These days, a hospital stay is reserved for patients undergoing surgery, or sometimes for a test that requires profound sedation or anaesthesia, like cardiac catheterization and transoesophageal echocardiography (one-day stay). Sometimes the child's heart condition is deemed serious enough to justify a hospital stay. The parents' presence is not only permitted, but also encouraged at all times. During hospitalization, several members of the medical staff, all with well-defined roles, will take care of the child. The mission of university hospital centres is to treat sick children, but also to train the next generation of caregivers. Thus, it is very likely that many students will be involved in the care, both during outpatient visits and during hospitalization.

Heart surgery

Pediatric heart surgery comprises all surgical procedures intended to correct, in most cases, heart defects in children. Except in the case of atrial septal defect and ventricular septal defect, heart surgery cannot reconstruct a heart as if it were completely normal. However, usually, after surgical repair, the heart will function almost normally.

The principle of surgical procedures is quite simple. It involves correcting or replacing a valve, repairing a heart segment or a partially obstructed vessel, closing an abnormal opening between the two heart chambers or positioning a conduit between two chambers that should be connected.

So-called palliative operations are performed in children with a heart defect that cannot be completely corrected. They aim to allow a better functioning of the heart, sometimes while waiting to be able to make a more complete repair. A typical example of a palliative operation is the creation of a shunt between the aorta and the pulmonary artery to increase the flow of blood to the lungs, i.e., the amount of deoxygenated blood that can collect oxygen from the lungs. As a result, the child will be less cyanotic (blue). The correction of univentricular defects, using the Glenn procedure and Fontan operation, is also considered palliative.

The purpose of corrective surgery as opposed to palliative surgery is to repair a defect completely. For example, a ventricular septal defect closure is considered corrective surgery: a patch is placed between the two ventricles to close the opening. In stenosis (a valve that is too small or too narrow), the valve can be repaired. Sometimes, if the valve has not developed properly, it may be necessary to replace it with an artificial valve (prosthesis). Another corrective operation is the repositioning of the aorta and pulmonary artery in a transposition of the great vessels (arterial switch), which allows normal circulation to be restored.

It should be noted that the majority of these procedures may leave a number of sequelae in the heart, such as an opening scar on the atrium or a ventricle, the presence of a patch of synthetic material inside the heart or a valve that is leaking. Those sequelae make it necessary to follow up throughout the patient’s life at varying intervals, even once the defect has been “repaired.”

When operations, palliative or not, do not require opening the heart, but are performed on the arteries or veins and outside the heart, the surgery is said to be closed-heart surgery. It requires opening the thorax under general anesthesia, but not the heart itself. When operations require access inside the heart, for example, to close an opening between the two ventricles, the heart must first be completely isolated from the circulation. This is called open-heart surgery. It requires cardiopulmonary bypass during which the function of the heart and the lungs is temporarily replaced by a heart-lung machine. The machine assumes both the pump function of the heart and the

function of oxygenating and removing carbon dioxide from the lungs. Although the idea is simple, cardiopulmonary bypass is extremely complex and requires the participation of a team of highly specialized healthcare professionals.

Usually, during cardiopulmonary bypass, and also sometimes during closed-heart surgery, it is essential to slow down the body's metabolism. This is achieved by using hypothermia, which consists in lowering the body temperature. This way, the heart will have to pump much less actively to support the body. Hypothermia will also protect the brain, which has a particularly high demand for oxygen at normal temperature. Once the operation is over, the patient is warmed up, cardiopulmonary bypass stopped, and the heart starts again spontaneously or, in some cases, with the aid of electrical stimulation.

Medication

Some medications are given specifically for a child's heart disease to help his heart perform its normal functions in the presence of a heart defect.

Diuretics are a type of medication commonly used in pediatrics. Their main purpose is to decrease the volume of circulating blood, forcing the kidney to excrete more fluid. This relieves the heart muscle. The main diuretic used is furosemide, also called Lasixtm. Because this medication causes the kidneys to excrete potassium, which is essential for the body, the doctor may sometimes prescribe another diuretic called aldactone to help reduce such excretion.

Another frequently used medication is **digoxin**, which main action is to strengthen the contraction of the heart, thus allowing the heart to pump enough blood through the body for vital functions. It is mainly used for defects involving a left-to-right shunt, such as ventricular septal defect, ductus arteriosus and atrioventricular septal defect. Since the child may regurgitate and vomit, it is best to give digoxin on an empty stomach. Indeed, when a child vomits, it is not possible to assess accurately how

much of the medication has been absorbed. In such case, it is safer not to give the child digoxin again and to wait for the next dose.

Also among the categories of medication given to children with heart disease are **vasodilators.** These medications encourage dilation of the blood vessels of the arteries in order to facilitate the passage of blood as well as the general functioning of the heart. This makes it easier for the organ to pump blood into the aorta. The most widely used medications in this category are captopril, enalapril, lisinopril and losartan. They will be prescribed for children with heart failure and weak heart muscle. Losartan is also used to prevent dilation of the aortic root in patients with Marfan's syndrome.

Some children need to be treated with anticoagulants to thin their blood. Most often they are given warfarin (or Coumadintm). These patients undergo regular monitoring and periodic blood tests to make sure their blood is thinned, but not too much. The doses of medication are adjusted according to the results of the blood test.

Some over-the-counter medications may also be used in children who have a heart problem. There is nothing wrong with giving acetaminophen (Tylenoltm or Tempratm) to a child with heart disease. If necessary, antibiotics may be prescribed for infections.

Nasal decongestants and cold syrups are also commonly administered. That being said, although they are very safe in general, these over-the-counter medications may still cause unwelcome and even dangerous side effects in some children with heart disease. They sometimes have the effect of increasing the strength of the contraction of the heart, causing some symptoms to worsen. It is therefore important to always consult a pharmacist before purchasing over-the-counter medications for a child with heart disease.

Pacemakers

Some children may need a pacemaker but their number is significantly lower than in adults. A pacemaker is a small electronic device used to restore a normal heart rhythm (Figure 29).

The principal indication for implanting a pacemaker is when the heart rhythm is too slow. For example, if a heart contracts only 40 times a minute, it will be difficult for it to pump enough blood through the arteries to meet the body's basic needs. The child will then feel weak and dizzy and his exercise tolerance will be greatly diminished. The slowing of the heart rate occurs either because the sinus node (the part of the heart that regulates the heart rhythm) ceases to send impulses or sends them too slowly, or because the impulses are blocked during their transmission from the atria to the ventricles. The vast majority of heart rhythms that are too slow occur as a result of heart surgery. This is because many surgeries require incisions or stitches in areas through which the major electrical conduction bundle passes. Unfortunately, it happens that the normal bundle is damaged. Other electrical cells will take over to keep the heart beating, but at a much lower rate.

Indications for the implantation of a pacemaker include a heart rhythm that is too slow for the child's age, excessive fatigue, or loss of consciousness. If this happens after surgery, it is wise to wait a week or two before making a decision as the heart's electrical circuits may sometimes start working again.

Figure 29 | Pacemaker

The pacemaker is a small device that contains a battery. Wires or electrodes bind the device to the heart muscle. This device regularly sends out electrical impulses by mimicking the nerve cells of the heart to allow the atria and ventricles to contract in sequence, thus restoring a normal heart rate.

Most pacemakers are permanently implanted under the skin and cannot be seen from the outside. Generally, one wire goes to the atria and another to the ventricles. Only one or the other of these electrodes may be needed, depending on the electrical circuit that is affected. Thus, when only the sinus node is affected and conduction between the atrium and the ventricles is not blocked, a single wire to the atria is needed. If there is a conduction block before reaching the ventricles, the pacemaker sends an impulse directly to the ventricles.

In older children, the electrodes are inserted into the subclavian vein, which passes directly under the clavicle. The electrodes follow this large vein towards the heart. The device is buried in a pocket under the skin under the clavicle or in the armpit. In smaller children, electrodes are connected directly to the heart because the veins are too small to insert the wires. The device is then usually placed in the abdomen at the level of the left flank.

There are also temporary pacemakers that the surgeon may place after heart surgery. The wires, attached to the heart muscle, come out through the skin and connect to a pacemaker. They are removed before leaving the hospital.

A pacemaker is a minicomputer capable of performing several functions. For instance, when the transmission of the current between the atrium and the ventricles is blocked, the pace-maker can sense the atria's contraction and transmit a signal to the ventricles to follow suit. It can thus adjust the rate of the heartbeats to the needs of the child (slow down when sleeping and accelerate when running). The functioning of the pacemaker can be analyzed at any time through the skin. It is also possible to transmit new instructions to it and adjust certain parameters such as the intensity of the current, the rate, the time between the stimulation of the atria and ventricles, etc.

In recent years, great advances have been made in pacemaker technology. More and more compact, some devices measure less than three centimetres from side to side and are less than one centimetre thick. Their battery life now ranges from 10 to 15 years. During an outpatient visit, the doctor will test the pacemaker and determine if it is time to replace the battery. The electrodes are usually permanent and only the battery needs to be changed in a relatively simple procedure. Remote monitoring systems are now available. The data transfer device, installed at home, communicates with the pacemaker and sends the information to software accessible by the healthcare team. If any abnormalities are detected, e.g., a low battery or a broken wire, an alert is automatically sent to the doctor.

A child with a pacemaker can go about all of his usual activities without any problem. However, it is recommended to avoid shocks to the device site and not to wear a cell phone directly on the device.

Implanted defibrillator

The implantation of an internal defibrillator may be necessary for some children. This sophisticated device can detect severe arrhythmia and send a

shock to the heart to stop it (Figure 30). This is actually the same treatment that is given to a person in cardiac arrest by sending an electric shock to the chest with an external defibrillator. Internal defibrillators are used in children who have had very serious arrhythmia or are thought to be at risk of having one.

Figure 30 | Implanted defibrillator

Artificial heart valves

In some cases, when one of the heart valves is severely malformed (too narrow, severe leak or both), it may be necessary to replace it completely if the surgeon feels that it is beyond repair. A replacement valve or prosthesis is then installed. Valvular prosthesis can be biological or mechanical. A biological valve is made from human tissue (allograft) or animal tissue (xenograft). The animal valves used are nearly always porcine valves; the prosthesis is then called a porcine biological prosthesis. The valve is fixed on a metal ring. Mechanical prostheses are completely artificial and can be installed as a replacement for any valve of the heart. However, they require the administration of medication (warfarin) to prevent the blood from clotting around the valve. These clots are dangerous because they can be

released into the circulation and obstruct very important small arteries, such as arteries in the brain.

Careful monitoring is required when taking warfarin. The level of anticoagulation should be kept within values determined by the doctor. If the blood is not thin enough, clots may form. If it is too thin, there is a risk of bleeding. The monitoring required to keep the effects of the medication under control is done through regular blood tests, but it is now possible to measure coagulation at home using a small, easy-to-use device (Coaguchecktm) (Figure 31), as is done for blood sugar in diabetes.

Mechanical valves are very durable and can even last a lifetime if the valve is large enough at the time of implantation. As for biological prostheses, they do not require the administration of warfarin, but their lifespan is much shorter. Accumulations of calcium deposits are often observed and prevent them from functioning properly. These calcium deposits will form more rapidly when the child is young.

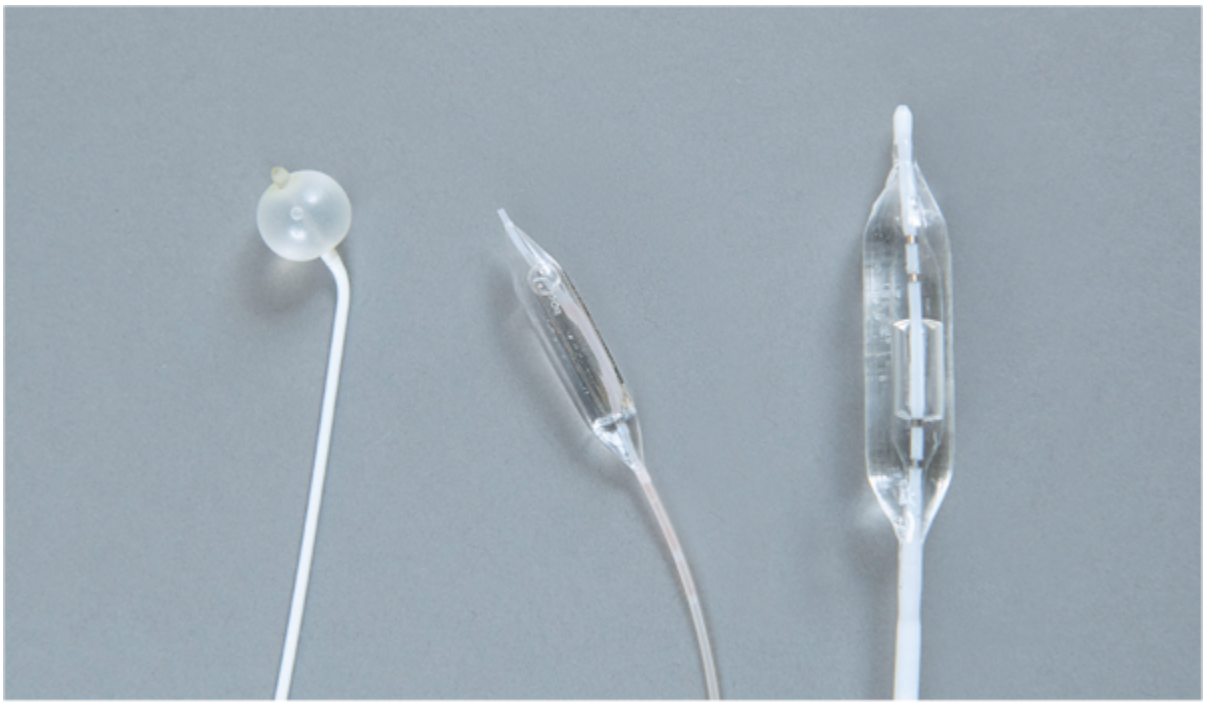
A child with a prosthetic valve can have a fairly normal active life. If he takes anticoagulants, he will need to be more careful when engaging in contact sports with risks of violent shocks that could cause bleeding under the skin or in the organs. Children who have had a biological or mechanical valve implanted also need to protect themselves against bacterial endocarditis, a heart infection often caused by bacteria in the mouth. They must therefore have good dental hygiene, brush their teeth well, floss, go to the dentist at least twice a year and take antibiotics one hour before some dental treatment.

Figure 31 | Coaguchek^{TM}

Interventional catheterization or heart procedures without a surgeon

Since the late 1980s it has become possible to correct many congenital heart defects without resorting to conventional surgery, that is to say without having to open the chest and the heart. This procedure is called interventional catheterization. It consists of introducing, through the veins or arteries, small special catheters (Figure 32) with which it is possible to dilate, cut and install prostheses. These catheters are miniaturized in order to enter the blood vessels, even those of very small babies. This procedure is done under sedation or general anesthesia and the child is hospitalized for only 24 hours in the majority of cases.

Figure 32 | Catheters used for interventional catheterization



From left to right: balloon catheter to create an atrial septal defect for a transposition of the great vessels (Rashkind septostomy); balloon catheter with blades to dilate a vessel; balloon catheter to dilate a larger vessel or valve.

Interventional catheters

Balloon catheters can be used to create an opening between the two atria (to facilitate the exchange of oxygen in a transposition of the great vessels), to dilate heart valves that are too narrow, especially the pulmonary valve and the aortic valve (aortic valve stenosis, pulmonary valve stenosis), or narrowed arteries such as the aorta (coarctation of the aorta) or pulmonary arteries (peripheral pulmonary stenosis). In older children, the success rate of artery dilation with balloon catheters may be improved by implanting a stent. It is a rigid wire mesh which, when implanted at the narrowing, maintains the vessel opened at the desired size. Stent placement is usually followed by treatment with aspirin for a minimum of six months.

Using certain catheters, it is also possible to cut some structures, in particular the atrial septum in order to allow blood from the pulmonary veins and vena cava to mix to ensure oxygenation in certain complex defects (tricuspid atresia, for example). This type of catheter has a small,

very sharp blade of about 1 cm in length on its tip which is opened with a mechanism when the catheter's top is in the left atrium. The catheter is then withdrawn quickly, and the blade cuts the septum. This device can also be used to open peripheral pulmonary arteries that are too small. The use of catheters that can deliver energy in the form of radio frequency makes it possible to open imperforate valves as in pulmonary atresia.

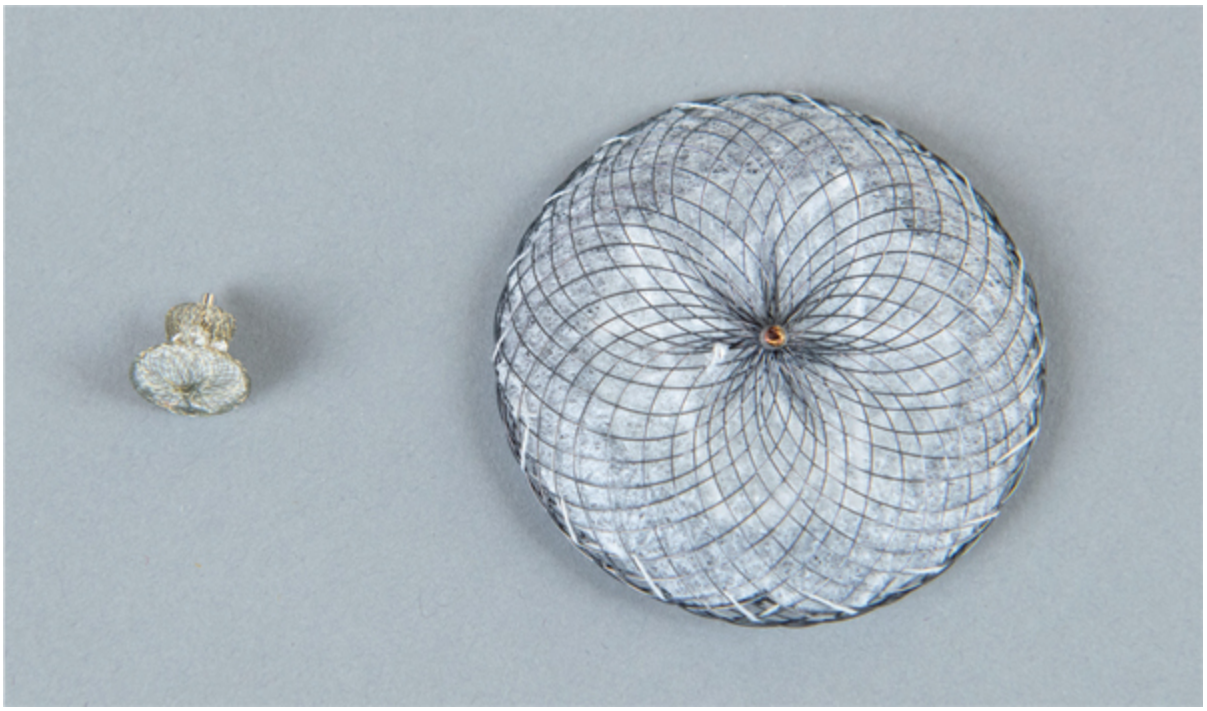
Endovascular prostheses

It is also possible to close certain abnormal openings using a variety of prostheses. Closure of a small ductus arteriosus is accomplished by means of small spring-like devices in most children. Those coils are covered with a filamentous mesh which causes the formation of clots thus blocking the ductus arteriosus. When the diameter of the ductus arteriosus is larger, an endovascular prosthesis is used. This type of prosthesis is made from various metal alloys. A wire of this alloy is woven into a very fine fabric shaped like a stopper flanked by two retention discs. In the central part of the stopper, a piece of waterproof fabric is sewn to the metal framework. The principle is simple: the prosthesis is anchored by the retention discs, while the central piece of fabric prevents the circulation of blood. This prosthesis bears the name of its inventor: Dr. Amplatz.

This type of prosthesis has found its major application in the closure of atrial septal defects (ASD) (Figure 33). In fact, this procedure is now most often done with a prosthesis. Less frequently, the prosthesis is used in the closure of muscular type ventricular septal defects.

The prosthesis is implanted under general anesthesia or under sedation and under fluoroscopic (X-ray) and echocardiographic (transesophageal echocardiogram) guidance. The child is hospitalized overnight and is usually discharged the next day after undergoing check-ups. He will need to take a small daily dose of aspirin for six months after the procedure.

Figure 33 | Endovascular prostheses



Interventional catheterization is a safe treatment. However, important complications may occur. The nature of these complications depends on the child condition and should be discussed in depth with the cardiologist responsible for carrying out the procedure. Suffice it to say that the risk of severe complications during a catheter procedure is no greater than with equivalent surgery. In fact, in the vast majority of cases, cardiac catheterization carries less risk, is less painful and poses fewer problems than surgery, while being just as effective.

There are many advantages to interventional catheterization: a brief hospital stay, no requirement for intensive care, no scar, considerably less anxiety in children and families and much lower cost for the health care system. The patient enters the hospital the day of the procedure and comes out the next day, sometimes even the same day. Nine times out of ten, he is cured of his defect. For some defects, the results are generally as good as those of open-heart surgery and sometimes even better. However, it is preferable to discuss the advantages and disadvantages of both techniques with the cardiologist in order to make an informed decision.

With some defects, such as pulmonary stenosis or aortic stenosis, the problems may recur after the procedure.

Heart transplantation in children

About 50 years ago, the first heart transplants in adults created enthusiasm all over. However, this enthusiasm was short-lived as the majority of transplant patients died fairly quickly following a rejection episode. Subsequently, there were fewer and fewer heart transplants worldwide. They finally resumed in greater numbers and this time with a better chance of survival, after very effective drugs against rejection became available. In pediatrics, things were slower to get started, but heart transplantation is now available to children and even newborns.

Generally speaking, heart transplantation is reserved for children with absolutely incurable heart disease, for example severe disease of the heart muscle (cardiomyopathy) or certain forms of inoperable heart defects.

Unfortunately, the number of donors is limited. A donor must be a child who is brain dead and whose still viable heart can be removed for transplantation. To determine which child will be transplanted, there is a relatively complex evaluation mechanism conducted by a multidisciplinary team. The following are assessed: the child's health status, both psychological and physical, the capacity of the family environment to provide the transplanted child with the necessary support which is relatively complex during the first years, and the overall psychological condition of the parents.

Survival after transplantation has improved, with five-year mortality after transplantation in the 15-20% range. However, be aware that the average lifespan of a transplanted heart is 10 to 20 years, sometimes more, depending on the child's age and health status. Some patients may need a second transplant.

In the first months following the operation, prolonged hospitalization followed by very frequent monitoring visits and multiple tests are necessary. Some of them, like myocardial biopsy, are unpleasant, but not painful for the child. Over the following years, this monitoring becomes less constraining and the child can lead a more normal life. He is even able

to exercise and play like other children his age, but he must take medication continuously.

Transplantation is not a panacea. While it can save the life of a child with a severe and incurable heart disease, it should always be seen as a last resort. The scarcity of donors is currently the main problem. Let us hope that current research efforts can help avoid occasional complications such as rejection, coronary obstructions, and drug-induced reactions. Other treatment solutions will also need to be developed, such as the use of hearts from other animal species or mechanical hearts.

It is important to say a word about organ donation. Obviously, if a child receives a heart transplant or any other organ transplant, it is because there was a donor, i.e., a person who, during his lifetime, agreed to donate organs (heart, liver, lungs, etc.) after death. Ideally, everyone should formalize this consent when renewing their health insurance card or by registering in the Registre de consentement au don d'organes et de tissus of the Régie de l'assurance maladie du Québec. Parents should also discuss the subject with their child when the child is old enough to understand what organ donation is.

The staff

In the course of the many contacts with the hospital, families will have the occasion to meet many members of the staff. The **pediatric cardiologist** is the one that takes charge of the child right from the first contact. He or she is the one who will do the follow-up continuously during outpatient visits but not necessarily the one who will see the child during hospitalizations or who will perform the cardiac catheterizations. At any time, if something is wrong, you should first contact the cardiologist. He or she is the person who knows the child and the family best.

The **heart surgeon** is the person who performs heart surgery. Not all children need surgery, but if surgery is required, the surgeon will be the one to do it. Surgeons are responsible for the ultimate decision to operate, the

surgical procedure and the postoperative care. Families usually have less contact with them than with the cardiologist, but in all cases the heart surgeon will meet with the child and the family before the operation. The surgeon will answer all questions related to surgery, including those relating to operative risks.

The **anesthesiologist** or anesthetist is responsible for putting the child to sleep before the operation and waking him up when it is finished. He or she is also actively involved in monitoring the child's vital signs during surgery. Usually, the anesthesiologist meets the child and the family during a preoperative visit in preparation for surgery.

Postoperative care is carried out by specialized doctors whose main role is to deal with all possible complications. Those are the **intensivists** (specialized doctors who work in intensive care). They are the ones who take care of the child while in intensive care after surgery.

The **respiratory therapist** is a specialized technician who works jointly with the anesthesiologist and the intensivist. The main role of the respiratory therapist is to watch the child's breathing, adjust the mechanical ventilator, and make sure that all the equipment needed to support the child's breathing is functioning properly.

The **perfusionist** is a technician specialized in the operation of the heart-lung machine during the operation. The sole responsibility of the perfusionist is to prepare the heart-lung machine and monitor its use during open-heart surgery.

Many **nurses** with various duties also take care of the child. There are nurses specialized in intensive care, operating room, hospital care, outpatient clinic, etc. Those are the people who take the child's vital signs, administer the medication, monitor the child closely, and notify the doctor at the slightest warning sign. They are also present in outpatient clinics and are often the first contact during appointments.

Several **specialized technicians** are also involved in the care of children. In particular, the radiology technician is responsible for taking the X-ray

images; the cardiology technician will record electrocardiograms, set up the Holter machine or provide the home ECG transmitter and perform the preliminary analysis of the Holter recording; the echocardiography technician is responsible for taking and ensuring the quality of the ultrasound images that the cardiologist will then interpret; hemodynamic technicians are present during cardiac catheterization to measure and record the pressures and oxygen levels in the blood, ensure the preparation of the catheters, their irrigation, the preparation of the laboratory equipment and other laboratory analysis.

The main role of **physiotherapists** in cardiology is to ensure good respiratory function and clear airways in children by having them do breathing exercises. They may also intervene if the child is hospitalized for a lung infection or if he needs physical rehabilitation (especially in infants).

The **occupational therapist** is a specialized professional who oversees the development of the child. Among other things, he or she helps to improve the feeding habits of children who may have developed an aversion to feeding after gavage.

The **psychologist** will see a child if there are signs of psychological distress before or after surgery or during delicate or difficult situations that bring distress to the child or his family.

Pharmacists are medication specialists. They advise doctors on choosing the right medication and the right dose and monitor possible interactions. In a children's hospital, their role is also to find solutions to divide the medications into smaller doses, which are not available on the market. Because pharmacists in pediatric hospitals are familiar with the medications prescribed for children with heart disease, they are sometimes consulted by other pharmacists to help them offer the right medications and doses.

Social workers guide parents toward services offered by the community and government resources. In addition, they offer individual, marital or family psychosocial follow-up in more stressful situations, such as hospitalization. They also guide the treatment team by informing them of

the reality of the family so that the objectives of the various interventions are adapted to their daily lives.

Nutritionists are nutrition specialists. When a child with heart disease needs a special diet (especially young children for whom special milk formulas are required), it is the nutritionist who guide the doctor and who participate in monitoring the diet and weight gain.

It should be noted that the staff operates in the setting of a teaching hospital and, by definition, the hospital is a place for the teaching of all specialties mentioned above. The children and their families will therefore also have to meet **students** from each of these specialties. In particular, they will have several contacts with residents specializing in cardiology or heart surgery. Those are physicians with a licence to practice medicine and who train under the supervision of an experienced doctor. Some have reached an advanced stage in their training as specialists and will be able to care for children almost independently.

Chapitre 6

Reactions to the diagnosis

Diagnosis

When a child is born with a heart defect, the impact on the parents and other family members is major. At the time of diagnosis, whether during pregnancy or at birth, parents may experience a lot of emotions: shock, sadness, anger, anxiety, etc. It should be remembered that all feelings are acceptable and understandable. Everyone must give themselves permission and time to digest the news.

> *"When I was 20 weeks pregnant, I learned that my unborn baby had a transposition of the great vessels. The news knocked me out. For a few days, I just cried. Eventually I pulled through and, with the support of my family and professionals, I was able to feel a little optimistic. I tried to enjoy the last months of my pregnancy."*
>
> Sophie, mother of Benjamin, 2 years old

The grieving process that follows the announcement of a congenital heart disease should not be underestimated. When they are expecting a baby, most parents have very specific expectations about the life that they wish to share with their child. A diagnosis of serious illness completely changes those expectations. Although the outcome may be positive, this is not the life the parents envisioned for their child and so there is some mourning.

Several researchers have analyzed the reactions of parents who learn that their child has a heart disease. They say they felt emotional ups and downs after hearing the news. Upon receiving the diagnosis, they report experiencing shock, disbelief and stress regardless of whether the diagnosis was made before or after birth.

There are many ways to go through the acceptance process. We recommend parents to ask a lot of questions to the medical staff. Those questions should be noted down as you go so as not to forget them during meetings. There are no bad questions. Although it is difficult, we recommend limiting Internet searches. The Internet comprises a lot of information that is not always true and which may cause more harm than good.

It is important to talk about what you are going through with trusted people around you. The strategy of trying to deal with all emotions on your own in order to protect others or because you believe “you can handle it” can quickly get out of hand.

You should know that at any time, if needed, you can receive professional help. Whether at the hospital or at the CLSC, professionals (social workers, psychologists, etc.) are there to support families through this ordeal. Sometimes, being able to confide in someone with some emotional distance may bring relief. Those professionals can also provide information on the various resources available for parents of sick children, such as financial assistance from the En Cœur Foundation as well as special government allowances.

Treatments and hospitalization

The treatment, surgery and hospitalization phase can generate a lot of emotions. A feeling of helplessness is not uncommon. During hospitalization, the role of the parents is not the same as when at home. It is recommended to be involved as much as possible in the care of the child. The child needs his parents, and his parents need him.

Another feeling experienced by many parents is guilt. Even though the medical team and those around them will say that the illness is not the parents' fault, it is sometimes difficult to accept.

> *"I would see my little baby with his scar, still getting pricked, and I would wonder what I did wrong during my pregnancy to cause the heart defect. It was not until later, when we were out of the critical period and had a chance to rest, that I really realized that it was not my fault. Doctors told me many times, but I had to be ready to hear and accept it."*
>
> Émilie, mother of Léa, 10 months old

Hospitalization and outpatient visits can also cause financial worries. Transportation, parking and meal costs add up. Often, there is also a loss of income, since at least one parent has to be absent from work. There are special government allowances that can help cover those expenses. If despite such allowances your financial situation remains difficult, the En Cœur Foundation can help.

What happens afterwards?

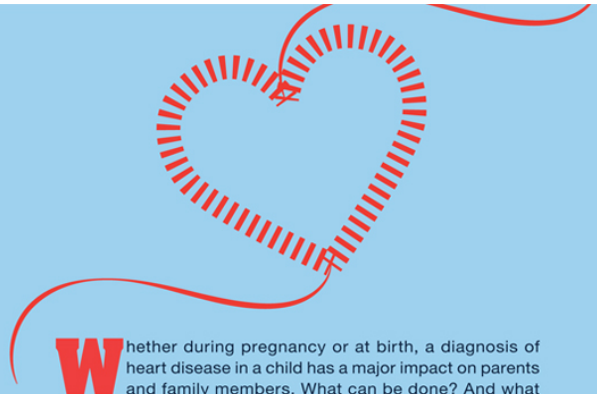
Just because the heart defect has been corrected, it does not mean that everything is resolved for all families. The psychosocial effects of the disease may even be more intense once the critical period has passed. You have to continue to take care of yourself as a parent as well as to watch over your relationship. Sometimes the reactions of each parent are very different which may cause tension in the couple. Maintaining good communication is essential. Again, do not hesitate to seek professional help if needed.

Parents are not the only ones affected by a child's heart disease. Depending on their age, several reactions are possible in siblings such as anger, jealousy or even regressive behaviour. Adjusting to a new baby is already a challenge for most children. When a baby is sick, the challenge is even

greater as it often means that the parents will be less present for the siblings. Several things may be considered in order to address this. First, it is important to have a routine. Do not hesitate to call on family members and friends with whom the child has a good relationship in order to maintain a sense of security. There are also resources such as books suitable for young children that discuss the diseases. The En Cœur Foundation has created a colouring book for the siblings which explains heart disease. Finally, professional support services are also intended for the family. They can be discussed with the medical team.

> *"The past year has not been easy for our family. As parents, we wanted so much to protect everyone—our sick child, his sister and his brother. We kind of forgot each other and we had some difficult times as a couple. Now that Ryan's condition is more stable, we have been able to take the time to talk and bond again. The other children also needed a lot of attention."*
>
> Adam, father of Ryan, 5 years old

That being said, studies have shown that many parents felt their bond grew stronger because of their child's heart disease. When preoccupied with procedures and hospitalization, extended family members were more likely to care for the other children at home, thus creating or strengthening lasting bonds.



Whether during pregnancy or at birth, a diagnosis of heart disease in a child has a major impact on parents and family members. What can be done? And what to except in the hours, days, months and years ahead?

The purpose of this book is to explain the heart to alleviate worry. It is intended for families with a child with heart disease, as well as all those who want to better understand the hearts of children. It is a remarkable reference, presenting varied heart defects, their physical, psychosocial and family implications, the therapeutic approaches to treat them, and the professionals called upon to intervene. It is also a great source of hope for better quality of life, as it bears witness to the medical care and the surgical techniques — less and less invasive — as well as the advancement of technology and resources available to affected families.

Written in collaboration with the **En Cœur Foundation,** this guide brings together the expertise of professionals who cares for and accompany children with heart disease and their families. This 3rd edition was written by Frédéric Dallaire, Pediatric Cardiologist in the Division of Pediatric Cardiology at the CIUSSSE-CHUS and Associate Professor in the Department of Pediatrics at the Université de Sherbrooke; Dr. Anne Fournier, Pediatric Cardiologist in the Division of Pediatric Cardiology at the CHU Sainte-Justine and Professor in the Department of Pediatrics at the Université de Montréal; and Dr. Claudia Renaud, Pediatric Cardiologist at the MUHC.