

Univentricular or biventricular repair in
HYPOPLASTIC LEFT HEART SYNDROME

Thesis Master Scientific Illustration

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Colophon

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**abbreviations and numbers, as for all images, are explained at the cover flap.*

PREFACE

I remember, as a child I dreamed of becoming a doctor. But as my passion for drawing and observing objects and nature started to grow and take over, I decided this was the one thing I wanted to take to the next level, despite my other interests. Although the Art studies in Gent increased my skills, the idea of being a contemporary artist did not respond to my goals, as I was forced more and more into the world of conceptual arts. That is when I decided to go into another direction.

The Master Scientific Illustration in Maastricht perfectly fits my interests. It allows me to apply my drawing talent to the second thing that always interested me: Medicine. My excitement for the profession of medical illustration never stopped growing since, as well as my fascination for the human body. After two years of exploring all aspects of the human body, in the end my attention was most caught by the beautiful design and complexity of the heart. The fascination started growing during the assignment in which I could study the tiny heart of a rat embryo. In contrast to that, I was even given the unique opportunity to investigate a massive elephant heart at the University Medical Center in Leiden (LUMC). It was a great experience working with this team in the research lab of the LUMC. During this week, I had the chance to talk to dr. M. Bartelings who allowed me to catch a glimpse of her work concerning postmortem examination on congenital heart defects. This really inspired me and in view of my Master Thesis, I could not let go off the idea that this would be an interesting and exciting subject area.

1

INTRODUCTION

1.1 PROJECT

Given my interest in the human heart and, more specifically, in congenital heart disease, I had the privilege to be introduced to cardiologist, dr. Derize Boshoff from the University Hospitals (UZ) Leuven, specialized in this field. As we discussed the possibility to cooperate in a graduation project, it became clear that there was still lack of good illustrations in some of these areas. Dr. Boshoff then introduced me to her colleagues' prof. dr. Marc Gewillig, head of paediatric and congenital cardiology UZ Leuven, and prof. dr. Filip Rega, adjunct-head of cardiac surgery UZ Leuven. These specialists recently began to cooperate on a new hybrid treatment strategy for children with hypoplastic left heart syndrome. Excitement started to grow as we discussed the subject and it became clear that this would be an interesting and challenging project in all aspects.

Hypoplastic left heart syndrome or 'HLHS' is a rare congenital heart disease in which the left-sided structures of the heart are underdeveloped or 'hypoplastic', including diminutive left ventricle, hypoplasia of the ascending aorta and ¹stenosis (narrowing) or ²atresia (absence) of the aortic and the bicuspid or mitral valve. The left side of the heart is unable to pump any blood to the body, which results in an overload of blood in the right ventricle as it has to maintain both pulmonary and systemic circulation. Usually there is a patent (open) interatrial connection, the oval foramen, present, and a large patent connection between the descending aorta and the pulmonary trunk, the arterial duct, supplies blood to the systemic circulation. The neonatal circulation in HLHS depends on these two connections.

However, within the first few days after birth, closure of the arterial duct starts and without intervention, there is usually a rapid deterioration with signs of low cardiac output and death within the first week of birth. Even if the arterial duct remains patent, death can be the result of progressive increase of pulmonary blood flow and decrease of systemic blood flow as the neonatal pulmonary resistance falls.

Until recently, the Norwood sequence remained the only treatment option for children with HLHS. This strategy can be described as a reconstruction consisting of three surgical procedures, performed to reconfigure the cardiovascular system to be as efficient as possible despite the lack of an adequate left ventricle. This is attained by creating a univentricular heart and a circulation in series, a so-called Fontan circulation, in which the heart no longer has to maintain both circulations, but only serves as a pumping chamber for the systemic circulation. However, this strategy contains major heart surgery and cardiopulmonary bypass with its associated risks in a neonate.

With a novel hybrid strategy, doctors created an alternative treatment for hypoplastic left heart syndrome with good early outcomes. It involves a less invasive procedure, where surgical techniques are combined with interventional cardiology techniques. The major advantage of this hybrid approach is the avoidance of early major heart surgery and bypass in high-risk patients. The goal is to create a stable and balanced circulation in the neonate, allowing the option to proceed to univentricular or biventricular repair in an older patient. However, the spectrum of conditions and the severity of the lesion make every case a challenge and different to approach. Therefore, the specialists classified all different cases into three groups. For each group a treatment strategy is proposed, but not completely definite, as in each case many other individual factors may demand another approach.

In this publication pathophysiologic, diagnostic and clinical aspects of the spectrum of HLHS will be systematically visualized and described, as there is no complete and uniform overview yet. As we focus on the clinical aspects of this syndrome, essential points of the different treatment strategies are illustrated, with the main focus on the new

hybrid approach, as it has not been visualized before. The illustrations will be an addition to the articles and publications written about this new treatment strategy. They should provide a good overview and a source of reference for paediatricians, surgeons, cardiologists or medical students. Furthermore, a schematic representation of the spectrum of HLHS is included in this publication. These illustrations will ultimately replace the basic elementary drawings of prof. dr. Gewillig and are now used as study material for medical students and for the explanatory information for patients.

“As I was looking for an effective way to show the different cases of this cardiac malformation, I wondered how I could visualize the specific information and details the target group demanded, despite the lack of good reference material.”

1.2 PREPARATION

In order to be able to make a good and structured overview with correct illustrations, perfect understanding of the subject is required. I was already familiar with the anatomy of the heart, as I had been studying it earlier during the Master Scientific Illustration. However, the condition of HLHS was still unknown to me. The internet and anatomy books provided poor information so I had to look for other sources. Remembering the work of dr. M. Bartelings in Leiden, I considered the added value of studying the development of the heart and the aetiology of HLHS. As we discussed this possibility, dr. M. Bartelings referred me to prof. dr. A.C. Gittenberger-de Groot, expertized in the biology of cardiovascular development and the pathogenesis of congenital heart malformations. Especially her doctorate work concerning HLHS was of great help to me. There were also several specimens with HLHS available at the University of Maastricht, so I dropped by frequently to study them, and also professional literature from the University and the specialists were of added value.

The specimens, however, did not describe the whole spectrum of HLHS but only the most severe cases, which still gave me an incomplete view on the complexity of the total spectrum. Therefore, communicating with the specialists in Leuven and reading their articles and case reports was of great importance to gain more insight into the pathology of the heart defect and the current treatment strategies.

1.3 PROCEDURES

HLHS is a rather rare congenital heart disease with only a few cases a year in Belgium. Considering that, these specific surgeries are not performed very often, which made it hard for me to make a planning. There are little occasions to see all cases of the spectrum, in the time frame of this Master Thesis. Despite this, I got the opportunity to attend a hybrid procedure on a newborn early in this project. Prof. dr. Rega guided me through the surgical part and prof. dr. Gewillig completed the procedure in the catheterization lab. It was great to experience the successful convergence of the two disciplines. During this procedure everything started to become clearer. I got a better view on the morphology of HLHS as the thorax was partly opened during cardiac surgery. However, the heart was not fully exposed. I immersed myself into the science of radiological imaging in order to gain more insight into the catheterization part of the procedure. The challenge for me now, was to reconstruct the heart based on the small part I had seen during cardiac surgery, and the radiological images made during cardiac catheterization to complement the rest.

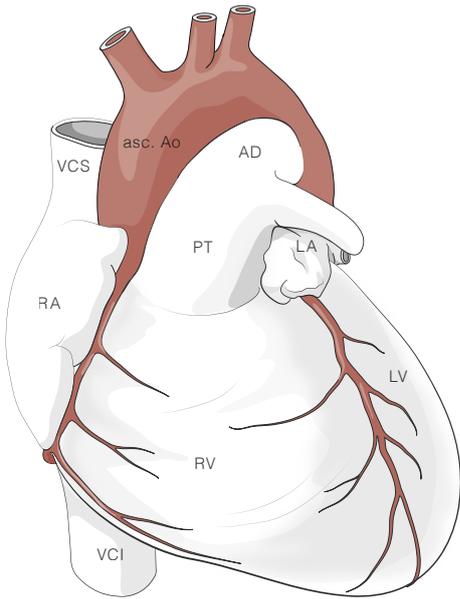


Figure 1
Anterior view of a normal heart of a neonate, before closure of the arterial duct.

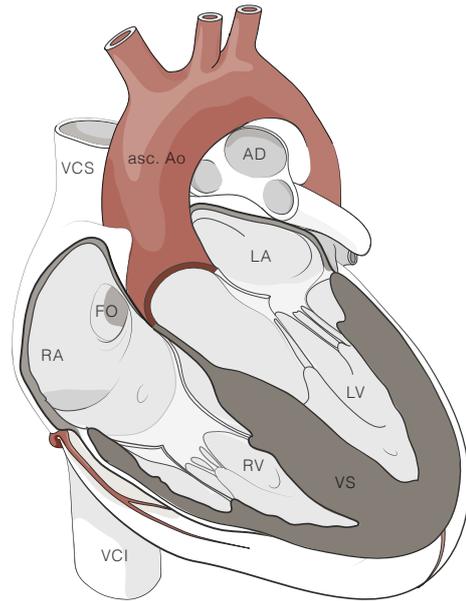


Figure 2
Cross-section of a normal heart of a neonate: four-chamber view.

1.4 BASIC ANATOMY

I wanted to start by giving the basic anatomy so this would not require further explanation. Thereby, I focus specifically on the aspects of the heart that are of importance in case of HLHS. Focal points are: the size of the ascending aorta and the aortic arch with placement of the coronary arteries, the shape of the heart, which is mostly determined by the size of the right (and left) ventricle and the right atrium, the thickness of the ventricular walls and septum, the relation of the arterial duct to the aorta, and the oval foramen. Furthermore the condition of the mitral and aortic valves during cardiac diastole and systole are of great importance, so superior views of the valves in these phases are added with marked aortic and mitral valves.

I have applied an approach of orientating the illustrations in this publication, where appropriate, as seen by the surgeon working in the operating room, though for most of the illustrations I have chosen an anatomical orientation. To clarify the various orientations, I included a set of axes, where needed, showing the directions of superior (S), inferior (I), anterior (A), posterior (P), left (L) and right (R).

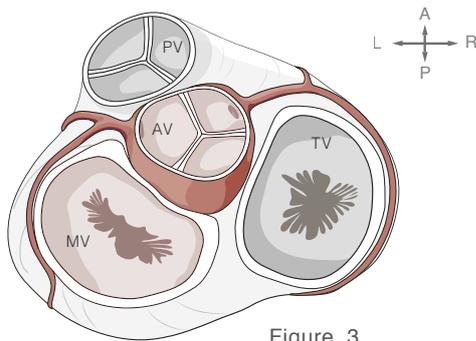


Figure 3

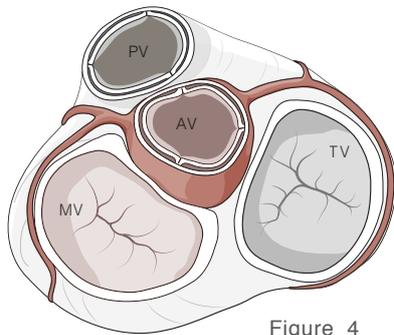


Figure 4

AORTA

Fig. 5 and 6 show the proportions of a normally developed aorta with its coronary arteries. Especially the size of the ascending aorta and the aortic arch should be noted, as well as the relation to the arterial duct, as it is of great importance when going to the spectrum of HLHS. The part of the aortic arch just distally to the origin of the left subclavian artery, at the site of the arterial duct, is called aortic isthmus. Figure 6 is showing the aorta in a postnatal situation when the arterial duct has already closed. As the condition of the arterial pathways are of great importance in case of HLHS, I chose to apply this view on the isolated aorta because only then it is fully exposed.

Figure 5

Anterior view of a normal aorta of a neonate, showing the relation with a large arterial duct.

Figure 6

Anterior view of a normal aorta, after complete closure of the arterial duct.

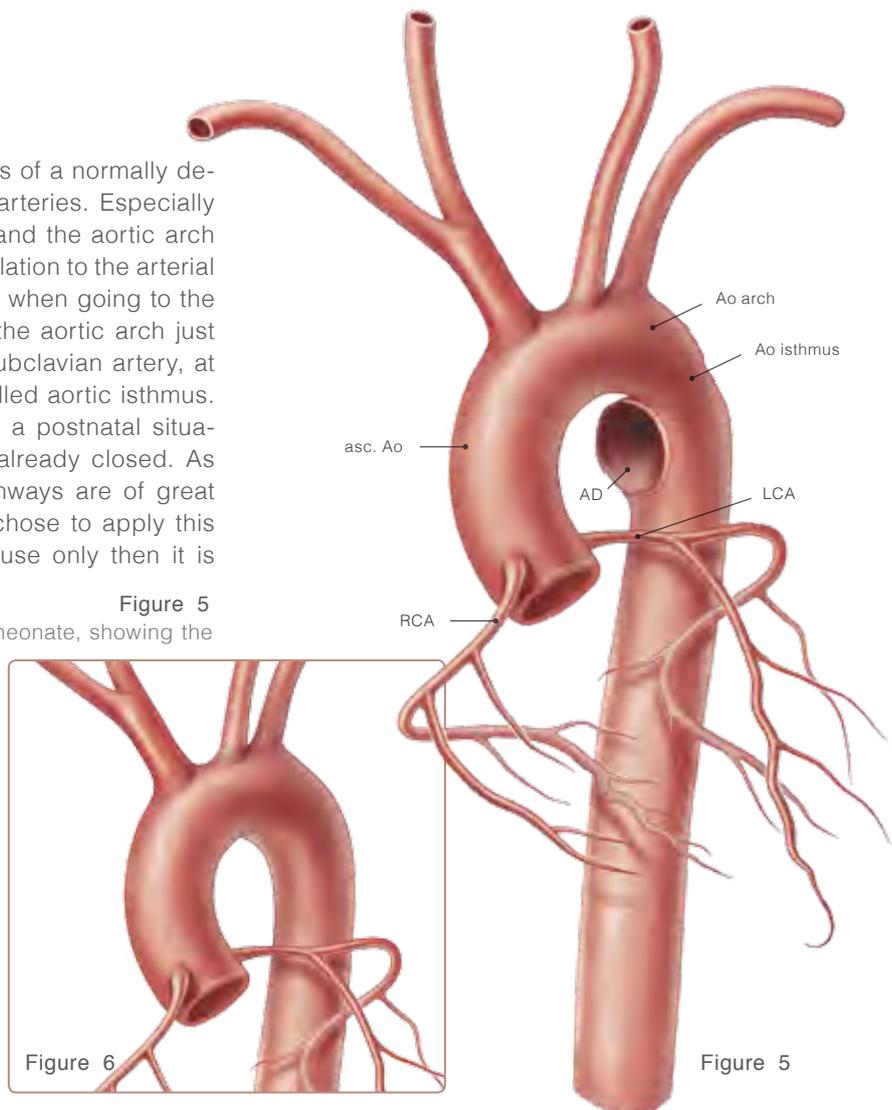


Figure 3-4

Superior view on the valves of a normal heart of a neonate: diastolic (fig. 3) and systolic (fig. 4) situation, showing the normal developed aortic, tricuspid and mitral valve.

DIASTOLE

The cardiac cycle consists of two phases: cardiac diastole and cardiac systole. During cardiac diastole, the ventricles relax and refill with blood. The pressure in the ventricles drops below the atrial pressure, causing the atrioventricular (tricuspid and mitral) valves to open and the blood to enter the ventricles. During this phase, the aortic and pulmonary valves remain closed.

SYSTOLE

Cardiac diastole is followed by cardiac systole. During this phase of the cardiac cycle, the left and right ventricles contract. The pressure in the ventricles increases again and when rising above the atrial pressure, the atrioventricular (tricuspid and mitral) valves close. When reaching the maximum pressure, the aortic and pulmonary valves open and the blood is ejected through these valves.

1.5 CIRCULATION

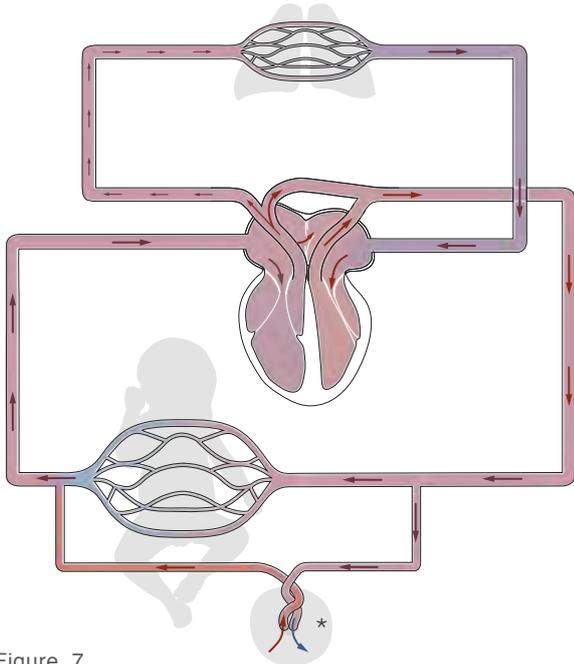


Figure 7
Schematic representation of a normal foetal circulation

Foetal circulation is characterized by the presence of the placenta, the arterial duct and the oval foramen. As the lungs are not functional before birth, the foetus receives its oxygen from the mother through the placenta (*) via the umbilical cord. In the foetus, only a small amount of blood is pumped into the lungs. Most of the blood bypasses the lungs through the oval foramen and the arterial duct.

Onset of breathing, expansion of the lungs and establishment of effective ventilation results in a decrease in pulmonary resistance and an increase of pulmonary blood flow. Closure of the arterial duct starts within the first few hours and permanent closure occurs in a few days. The increased pulmonary flow causes a rise in left atrial pressure, which results in functional closure of the oval foramen. Removal of the placenta, closure of the arterial duct and the oval foramen, and the increase of pulmonary flow cause a dramatic change in the cardiac flow pattern.

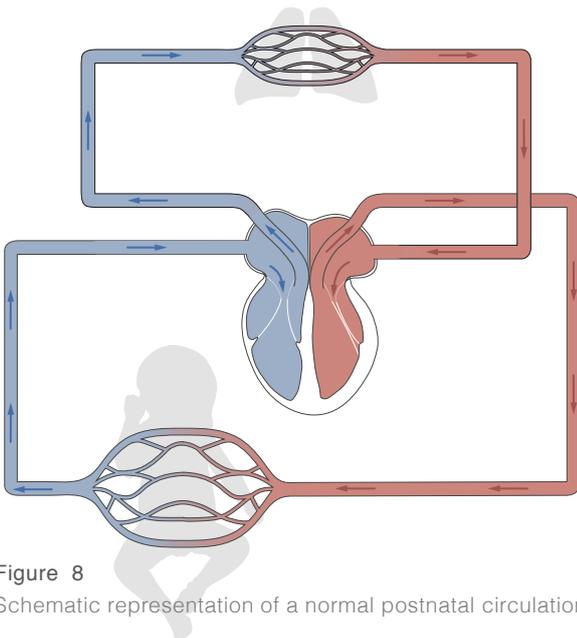


Figure 8
Schematic representation of a normal postnatal circulation



Method: To have a better overview of the normal and abnormal anatomy and function of the heart, understanding of the foetal and postnatal circulation is important. I tried to simplify the circulations in such a way that I could clarify the circulation in case of HLHS easily later on, as well as the Fontan circulation. The most important aspects I had to take into account were the left and right ventricles and outflow tracts, the oval foramen and arterial duct, and the balance of the pulmonary and systemic circulations.

Therefore, I decided to merge the superior and inferior caval veins in order to be able to show the systemic circulation in a more schematic way, to clarify it instantly. The illustrations of the hearts are based on the elementary drawings of the cardiologist (see next chapter), to have the exact proportions of the left and right ventricle with the correct thickness of the cardiac walls and septum.

I made the illustrations in Adobe Illustrator to create a clean and clear scheme. The use of colour and gradients was effective to show the degree of saturation of the blood in the heart and circulation. The arrows are showing the direction of blood flow.

2.1 THE SPECTRUM OF HLHS

To have a better and more structured overview of the complete spectrum of conditions that may occur, the specialists in Leuven made a classification, mostly determined by the size of the left heart. All cases can be divided into three groups, going from a slightly smaller than normal to a severely hypoplastic left heart. The first group is not called 'HLHS' because of the adequate sized left ventricle, aorta and valves. Only the isthmus might get too narrow (borderline sized) to sustain optimal systemic flow.

The third group 'HLHS' has two variants. Severe HLHS is probably the result of limited flow during development secondary to a primary abnormality (atresia in this case) of either the left ventricular outflow (aortic valve) or inflow (mitral valve). For each group, the most important aspects are systematically described and visualized.

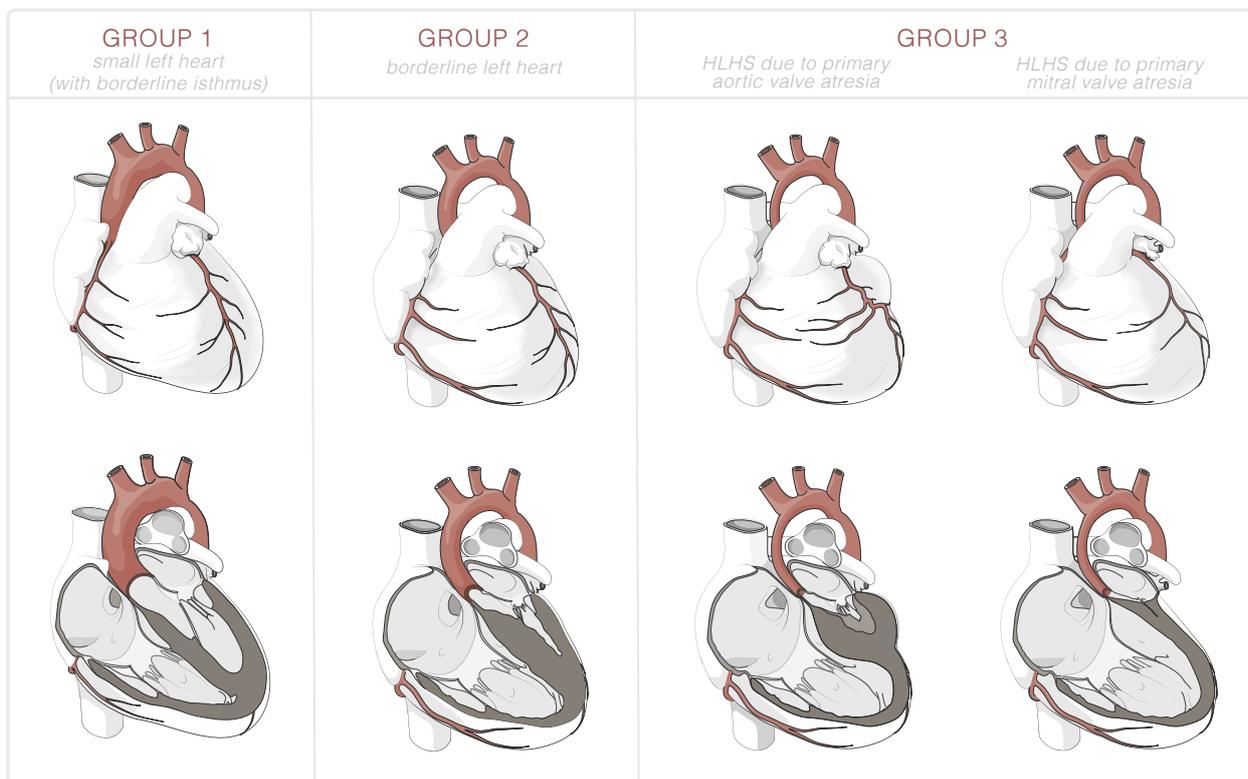
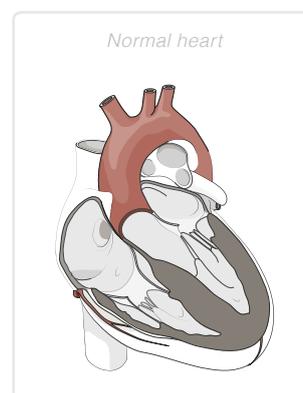


Figure 9

Overview of the different conditions of HLHS.

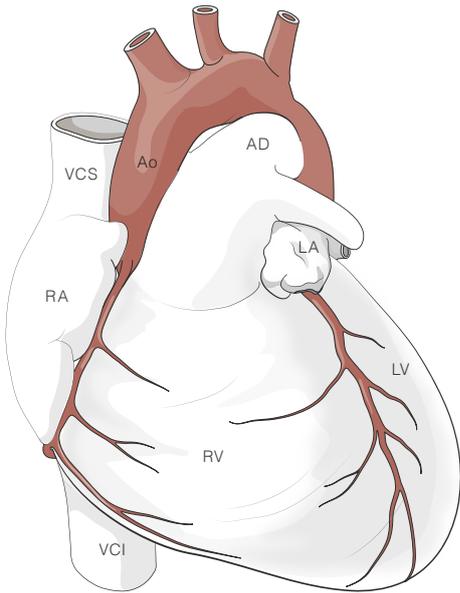


Figure 10
Anterior view of a heart of a neonate with a small left heart. The left sided structures are only slightly decreased in size.

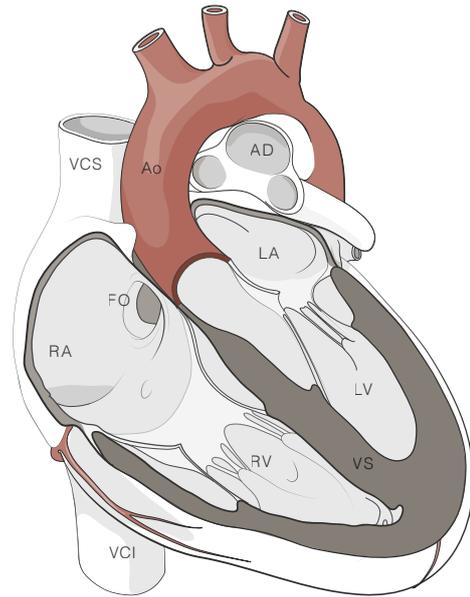


Figure 11
Cross-section of a heart of a neonate with a small left heart: four-chamber view. The left sided structures are only slightly decreased in size.

GROUP 1

Small left heart and borderline isthmus

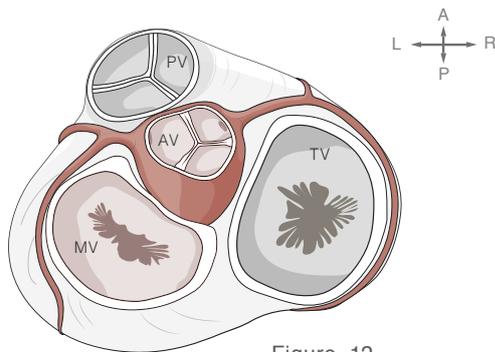


Figure 12

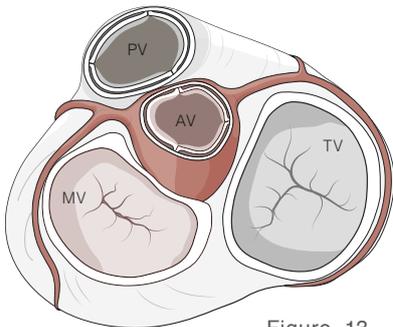


Figure 13

For this group the left sided structures of the heart are only slightly decreased in size and treatment is usually not necessary.

VENTRICLES AND VALVES

The aortic valve is decreased in size, as well as the left ventricle and mitral valve, and there is still an adequate amount of blood entering the left ventricle to be pumped out through the left ventricular outflow tract. The right ventricle is slightly enlarged, as well as the tricuspid and pulmonary valves. The ventricular walls and septum have a normal thickness.

Figure 12-13

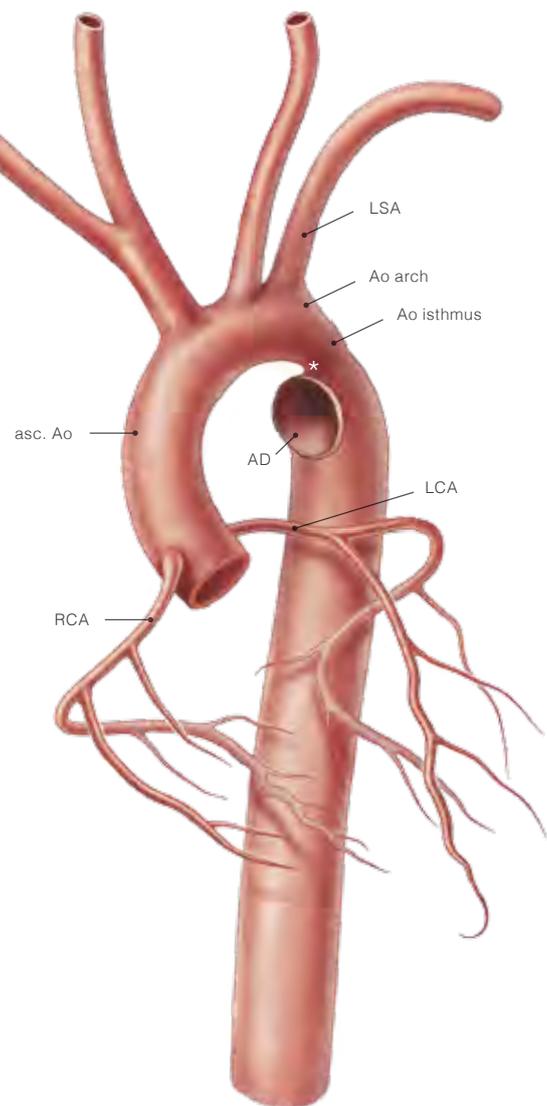
Superior view on the valves of a neonate with a small left heart: diastolic (fig. 12) and systolic (fig. 13) situation, with the aortic and the mitral valve both smaller than normal.

AORTA AND VALVE

The ascending aorta is smaller than normal and gives origin to normally positioned coronary arteries. The aorta comes together with a large arterial duct, distally to the left subclavian artery. The isthmus is narrower and may be too small after closure of the arterial duct : this slight narrowing of the aortic arch is called borderline ⁴coarctation (*).

Figure 14

Anterior view of the aorta of a neonate with a small left heart, showing the relation with a large arterial duct. The ascending aorta is smaller than normal.



Method: While studying and communicating with the specialists, it became clear to me that the visualisation of the spectrum of this cardiac malformation would be a challenging task. In an attempt to find an effective system to illustrate the anatomy in HLHS, I decided it had to be accurate and broad to be useful clinically, but also clear and consistent. In response to that, I tried out different techniques, like pencil or watercolour, to clarify the focal points of the spectrum. However in the end, a clean drawing in Adobe Illustrator with some local additions in Adobe Photoshop seemed the best solution to me as it allowed me to show only the essence without distracting the viewer with an overload of details. In addition, it gave me the ability to easily make adjustments, which was an advantage, as I had to make the drawings mostly based on the descriptions of the cardiologist and the cardiac surgeon.

For the illustrations of the anterior views of the hearts and the cross sections I decided to make line drawings in Adobe Illustrator. The addition of various tones of grey allowed me to suggest some three dimensionality but also to emphasize a certain area or aspect of the heart, like the thickness of the cardiac wall and septum.

The same way, I built up the illustrations of the superior views of the valves. For these images, the use of a transparent coloured field was effective to emphasize the condition of the aortic and mitral valves for every case of the spectrum. When illustrating the different conditions of the aorta, with the focus mostly on the ascending aorta and aortic arch, I decided to make an easy adjustable line drawing in Adobe Illustrator and further colour it in Adobe Photoshop in order to create a more detailed and three dimensional image.

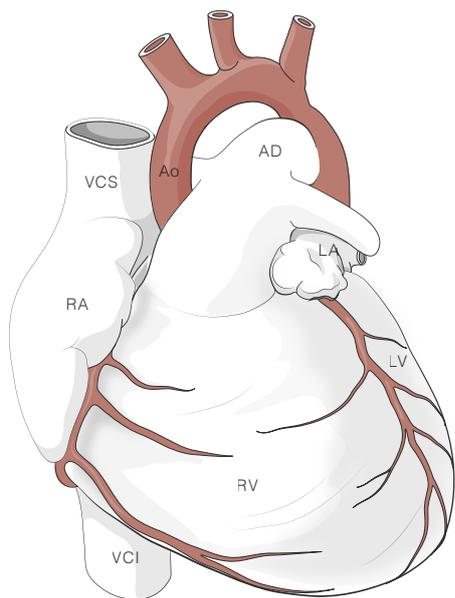


Figure 15
Anterior view of a heart of a neonate with borderline left heart.

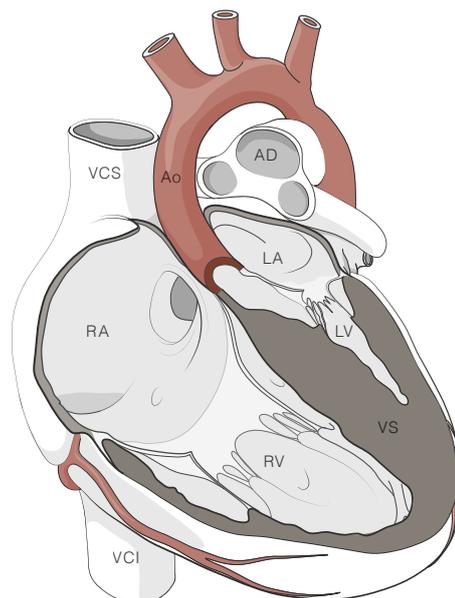


Figure 16
Cross-section of a heart of a neonate with borderline left heart: four-chamber view.

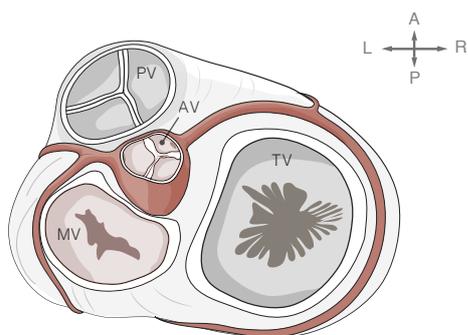


Figure 17

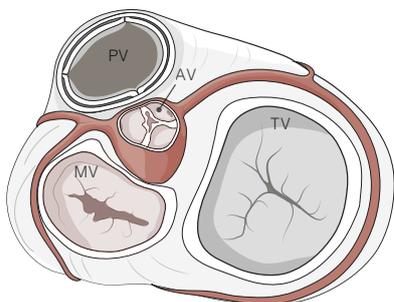


Figure 18

GROUP 2 *Borderline*

This central group of small left hearts is also called the 'borderline' group. The left sided structures of the heart are on the edge (the border) of being too small to sustain systemic flow.

VENTRICLES AND VALVES

The left ventricle is diminutive, frequently due to the restricted flow into the ventricle, which is (frequently) caused by a restrictive oval foramen. The right ventricular output is increased and often the right ventricle is enlarged, as well as are the tricuspid and the pulmonary valve. The aortic and mitral valves are small. Sometimes the aortic valve is stenotic (narrowed), which results in high pressure and thickened ventricular walls.

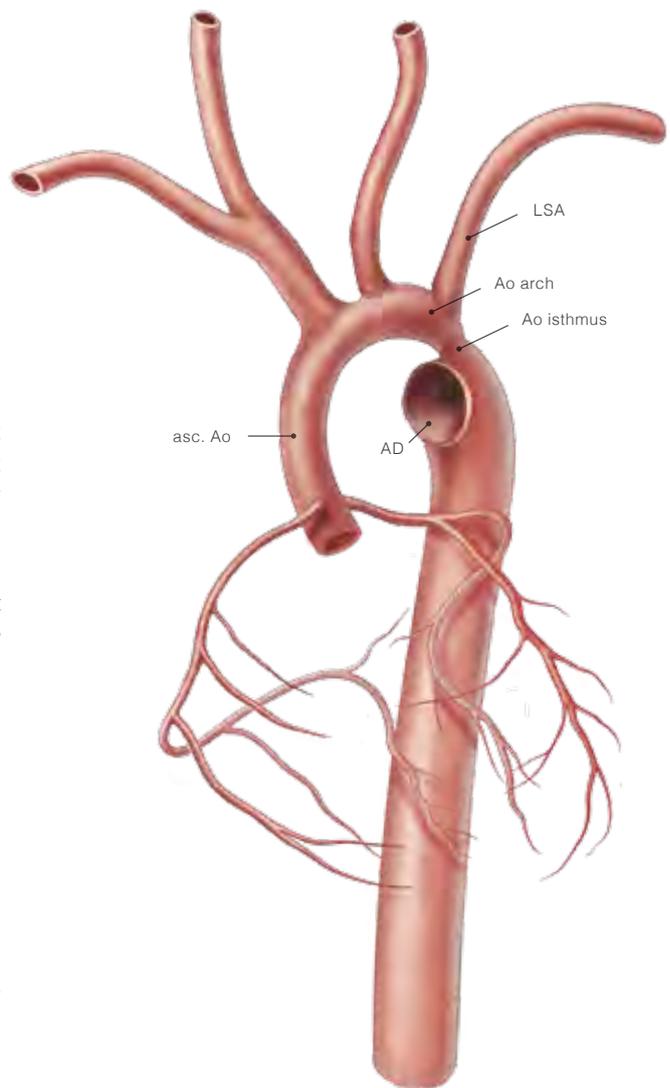
Figure 17-18
Superior view on the valves of a neonate with borderline left heart: diastolic (fig. 17) and systolic (fig. 18) situation, showing the aortic and mitral valves, being both small. The aortic valve may be stenotic.

AORTA AND VALVE

The aortic valve is small and may be stenotic, allowing a severely restricted flow into the aorta. The aorta is medium sized, with normally positioned coronary arteries, marking the borderline sized left ventricle. There is a large arterial duct present that comes together with the aorta beyond the narrow isthmus. There often is some retrograde filling of the distal arch.

Figure 19

Anterior view of the aorta of a neonate with borderline left heart, showing a narrow ascending aorta, a small isthmus and a large arterial duct.



RETROGRADE AORTIC ARCH

In a normal situation, blood flows from the left ventricle into the aorta and coronary arteries. There is a normal antegrade filling of the subclavian and common carotid arteries.

In case of aortic valve stenosis or atresia, there is a severely restricted flow into the left ventricular outflow tract, or it is completely blocked. These cases have a noticeable 'retrograde aortic arch', due to the retrograde filling of the aorta and its branches, into the coronary arteries. If the arterial duct closes, there no longer is blood supply to the body.

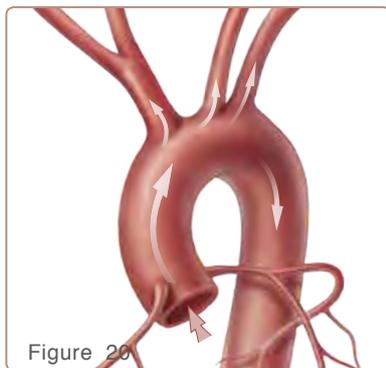


Figure 20

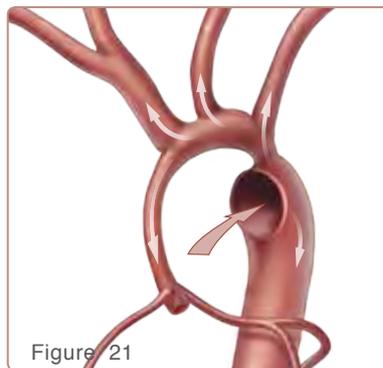


Figure 21

Figure 20

Anterior view of a normal aorta and arch, showing a normal antegrade filling of the aorta and branches.

Figure 21

Anterior view of a hypoplastic aorta and arch, showing a retrograde filling of the aorta and branches.

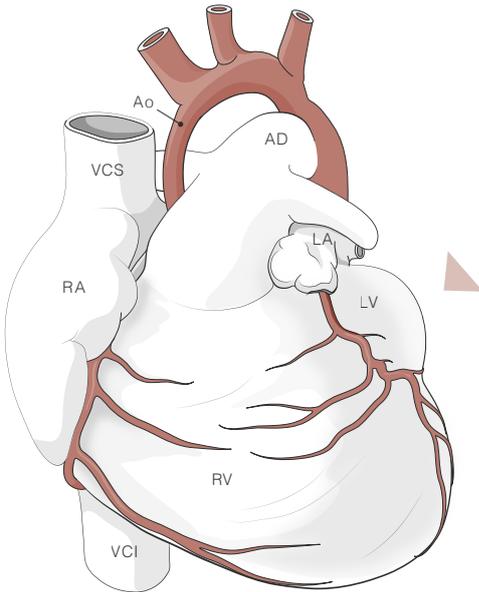


Figure 22
Anterior view on a heart of a neonate with HLHS. The left sided structures are severely hypoplastic.

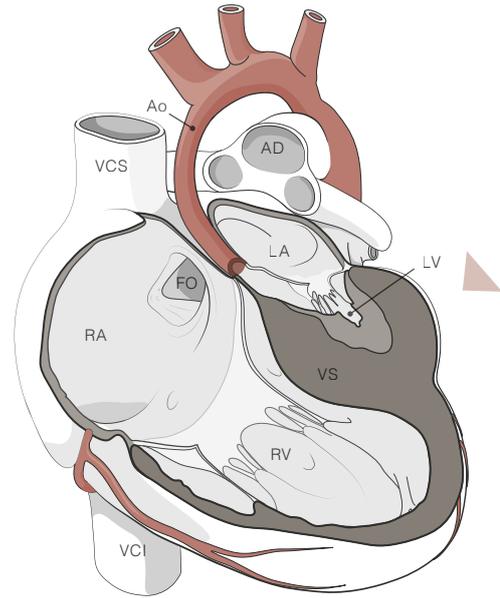


Figure 23
Cross-section of a heart of a neonate with HLHS: four-chamber view. The left sided structures are severely hypoplastic.

This is the most severe group called HLHS. The heart is enlarged and its shape is mostly determined by the large right ventricle. The aortic valve is usually atretic, the mitral valve small and stenotic, sometimes atretic.

GROUP 3a

HLHS due to primary aortic atresia

VENTRICLES AND VALVES

The aortic valve is atretic, the mitral valve is small, dysplastic and stenotic. There is a severely restricted flow into the left ventricle. Due to the atretic aortic valve, the blood is captured, which results in high pressure in the left ventricle. This causes the left ventricle to be round with thickened walls and a variable degree of thickening of the innermost lining of the heart chambers (endocardial fibro-elastosis). There is a marked bulging of the heart muscle or myocard (triangle). The right ventricle is usually hypertrophied with increased cavity size. The tricuspid and pulmonary valves are larger than normal and tricuspid incompetence is often present. There is a large pulmonary artery and a large right atrium.

Figure 24-25

Superior view on the valves of a neonate with HLHS: diastolic (fig. 24) and systolic (fig. 25) situation, showing an atretic aortic and a stenotic mitral valve.

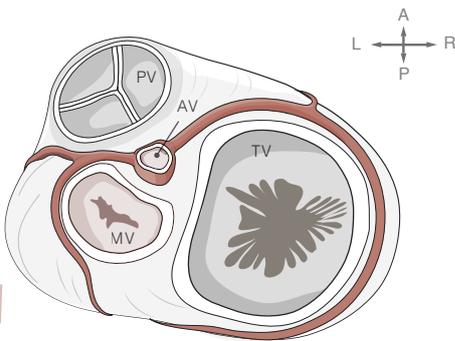


Figure 24

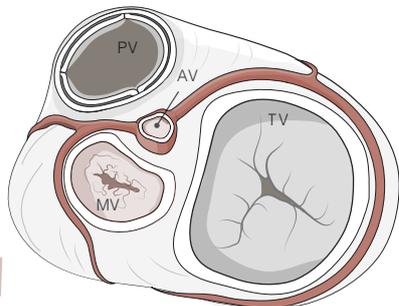


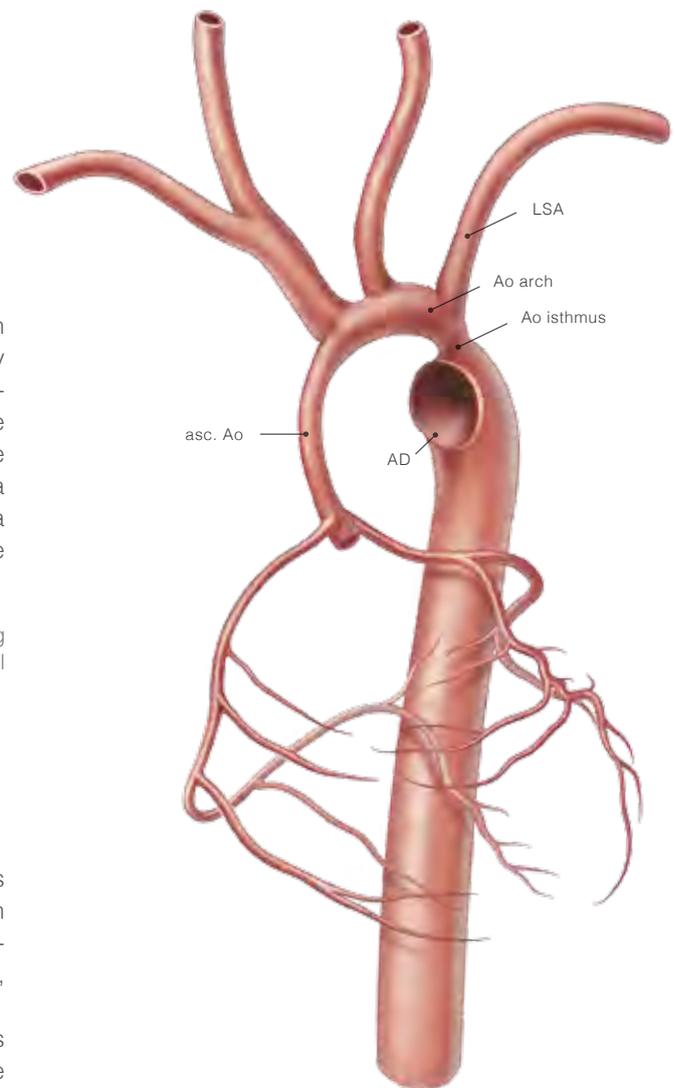
Figure 25

AORTA AND VALVE

The narrow ascending aorta, which gives origin to normal positioned coronary arteries, only serves as a main coronary artery with retrograde flow. More distally, from the innominate artery, the aortic arch widens and (beyond the left subclavian artery) comes together with a large patent arterial duct, which serves as a conduit to supply blood from the right ventricle into the descending aorta.

Figure 26

Anterior view of the aorta of a neonate with HLHS, showing a narrow, severely hypoplastic, ascending aorta, a small isthmus and a large arterial duct.



ENDOCARDIAL THICKENING AND OVAL FORAMEN

Due to the pressure in the left ventricle, there is a variable degree of endocardial thickening with fibroelastic tissue, also called 'endocardial fibroelastosis' or 'EFE'. This is a white scar-like tissue, further restricting the heart's pumping motion. A patent oval foramen is usually present and its valve is herniating into the right atrium, due to the pressure on the left side. Sometimes the valve is perforated (fenestrated) as well.

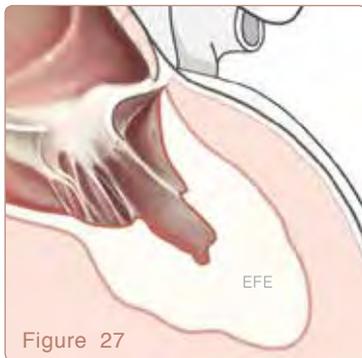


Figure 27



Figure 27

Left ventricle, cross-section: showing a white layer of endocardial thickening with fibroelastic tissue (EFE).

Figure 28

Right atrium, cross-section: showing the herniated valve of the foramen ovale into the large right atrium

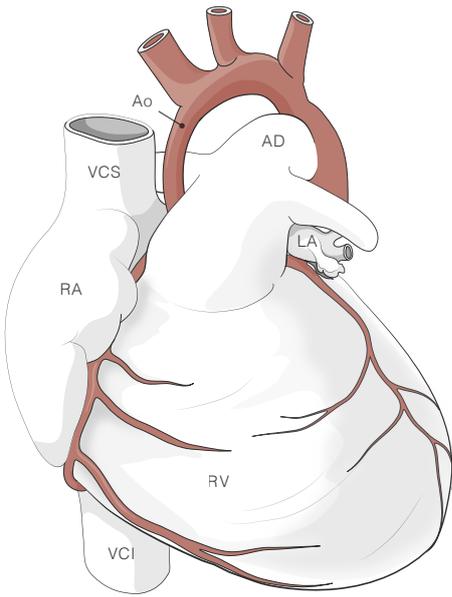


Figure 29
Anterior view on a heart of a neonate with HLHS. The left sided structures are severely hypoplastic.

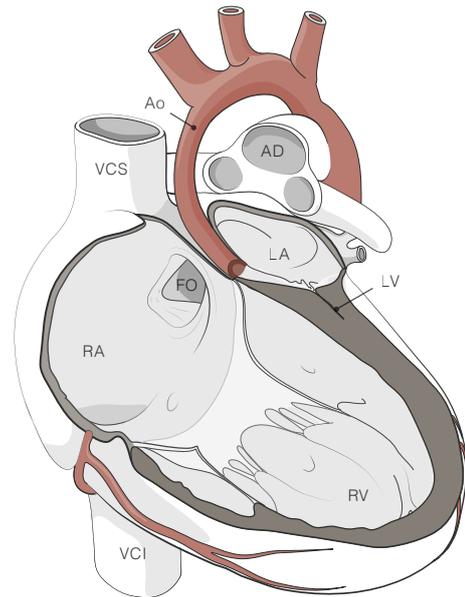


Figure 30
Cross-section of a heart of a neonate with HLHS: four-chamber view. The left sided structures are severely hypoplastic.

GROUP 3b

HLHS due to primary mitral atresia

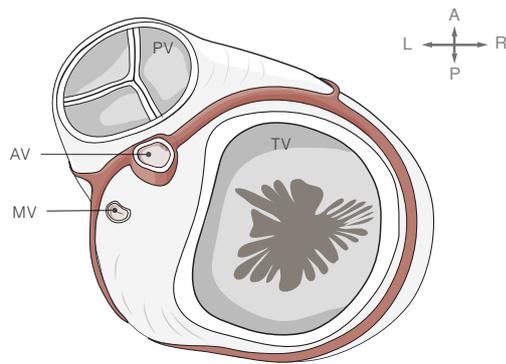


Figure 31

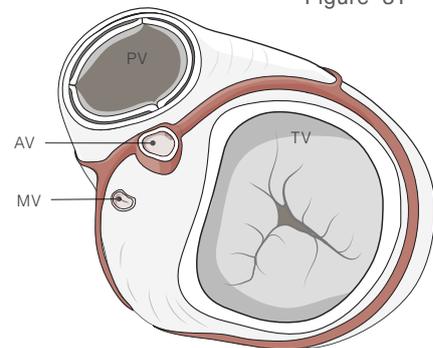


Figure 32

VENTRICLES AND VALVES

In case of aortic and mitral valve atresia, the left ventricle cavity is slitlike and often not visible and the wall almost nonexistent. The shape of the heart is only determined by the large right ventricle and atrium. Both valves are completely closed. Therefore, the blood can not enter the left ventricle, so it is shunted directly into the right ventricle through the oval foramen. The right ventricle is usually hypertrophied with increased cavity size. The tricuspid and the pulmonary valve are larger than normal and tricuspid incompetence is often present. There is a large pulmonary artery and a large right atrium.

Figure 31-32

Superior view on the valves of a neonate with HLHS: diastolic (fig. 31) and systolic (fig. 32) situation, showing the aortic and mitral valve, being both atretic. The pulmonary and tricuspid valve are larger than normal.

AORTA AND VALVE

The narrow ascending aorta, which gives origin to normally positioned coronary arteries, only serves as a main coronary artery. More distally, from the innominate artery, the aortic arch widens and (distally from the left subclavian artery) comes together with a large patent arterial duct, which serves as a conduit to supply blood from the right ventricle into the descending aorta. There is a retrograde filling of the aortic arch and ascending aorta, into the coronary arteries.

Figure 33

Anterior view of the aorta of a neonate with HLHS, showing a narrow, severely hypoplastic, ascending aorta, a small isthmus and a large arterial duct.

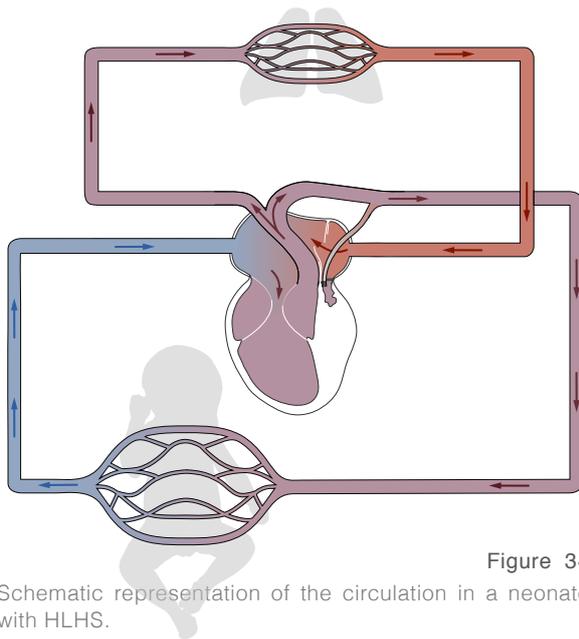
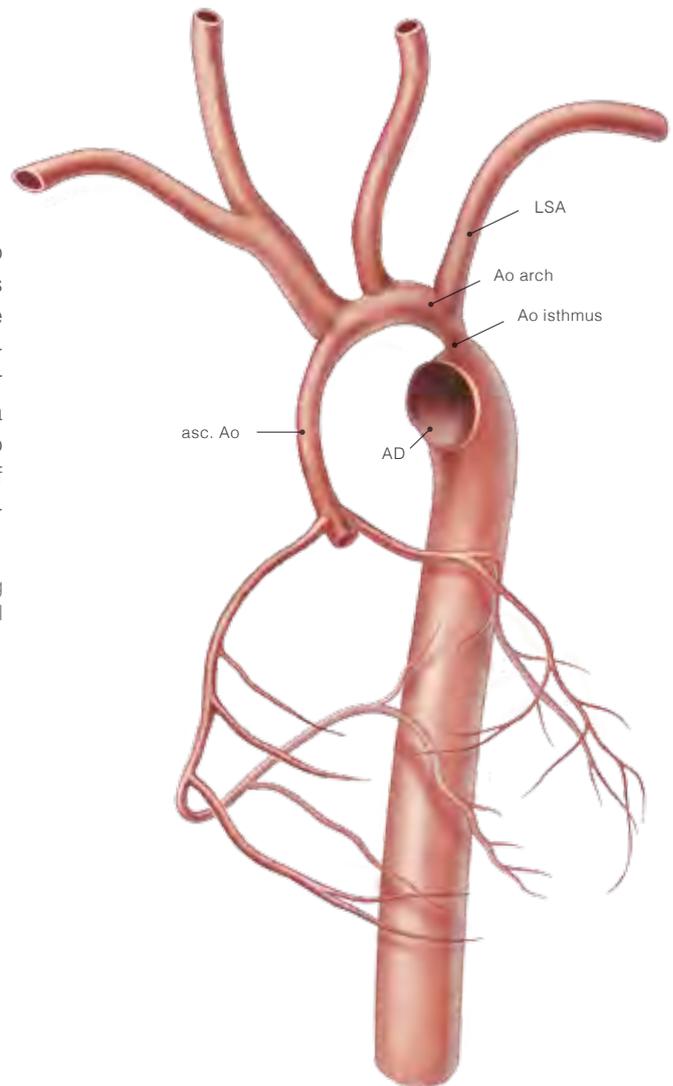


Figure 34

Schematic representation of the circulation in a neonate with HLHS.

CIRCULATION IN HLHS

The neonatal circulation in HLHS depends on the presence of the arterial duct and the oval foramen. The left ventricle and its outflow tract are too small, so blood must pass through the oval foramen. After entering the right ventricle it is pumped out through the pulmonary trunk. From there it partly continues on to the lungs, and partly supplies blood to the systemic circulation via the arterial duct. There is a mixture of systemic and pulmonary venous blood, so saturation is lower than normal. As the arterial duct and the oval foramen start to close after birth, it is clear that without intervention chances of survival are nihil.

2.2 DIAGNOSING HLHS AND INITIAL EXPLANATION

DIAGNOSING HLHS

HLHS can be diagnosed on foetal echocardiograms. The advantage of early diagnosis of this cardiac malformation is that delivering such a baby can be planned at an equipped hospital and intervention for stabilization of the neonate can be started immediately after birth. Thereby, severe shock is avoided and chances for a good outcome are improved. Neonatal echocardiography can give a lot of information about the severity of the lesion. It can give information about the size of the ventricles and outflow tracts, the condition of the valves, and the size of the atrial septal defect and the arterial duct.

During cardiac catheterization the exact size of the aorta and aortic arch can be outlined with contrast injection. Furthermore, it gives information about the cardiopulmonary function and anatomy. However, due to the risk in an unstable neonate, it is not used as an initial method for diagnosing HLHS. Later in life, it is useful to evaluate the function and anatomy of the heart when planning later stages of treatment and as part of the hybrid procedure.

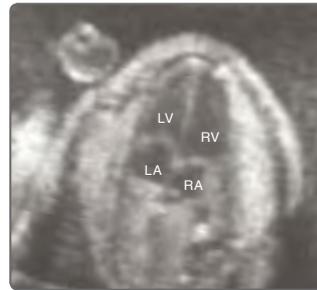


Figure 35
Echocardiography

Foetal echocardiography showing a normal physiology.



Figure 36
Echocardiography

Foetal echocardiography showing a small and hypertrophic left ventricle.

INITIAL EXPLANATION

To make an openingsstart of the description of the child's condition, prof. dr. Gewillig uses his 'famous' drawings to clarify the problem. The schematic drawings demonstrate a more basic approach of the heart, showing only the essence the parents need or want to know.

To optimize these elementary drawings, I made new and more elaborated versions, with more attention to details. Thereby not changing the concept but only making it more clear and precise as these illustrations are used as study material for medical students as well.

Method: Like for most of those parents, these drawings of prof. dr. Gewillig were my first acquaintance with the spectrum of this cardiac malformation. This approach of the heart is showing the right and left outflow vessels reflected to the apex of the heart. This creates a clear view on the atrial septum and foramen ovale. As we agreed the drawings could use an 'update', I looked for a solution to improve them without losing the concept. I decided to define the shape of the heart, and the thickness of the cardiac walls and septum. Further, I used the exact proportions, given by the specialists, to define the size of the aorta and pulmonary trunk, and the proportions of the left and right ventricles and atria.





Figure 37

Schaal ± 1:1

Figure 38

Schaal ± 1:1,5

Figure 39

Schaal ± 1:3

Figure 37-39

Figure 37: Normal foetal heart, Figure 38: Normal neonatal heart (1 day old), Figure 39: Normal mature heart. (as approached by prof. dr. Gewillig)

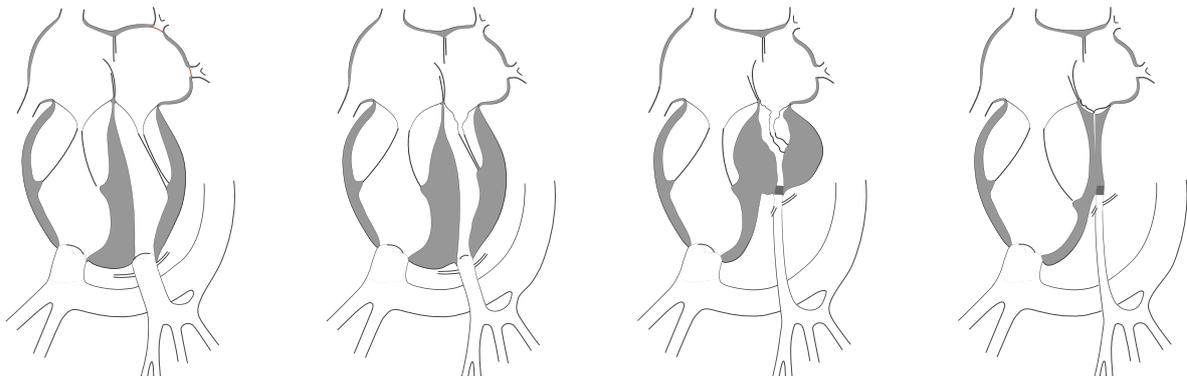


Figure 40

Figure 41

Figure 42

Figure 43

Schaal ± 1:1,5

Figure 40-43

Figure 40: Heart of a neonate with a small left heart and borderline isthmus (group 1), Figure 41: Heart of a neonate with borderline left heart (group 2), Figure 42: Heart of a neonate with HLHS due to primary aortic atresia (group 3a), Figure 43: Heart of a neonate with HLHS due to primary mitral atresia (group 3b). (as approached by prof. dr. Gewillig)

3

TREATMENT

3.1 NORWOOD SEQUENCE

Until recently, the Norwood sequence remained the only option for children with HLHS. This surgical correction involves a reconstruction of the arterial pathways and consists of three surgical procedures: The Norwood procedure (stage I), the Bidirectional Glenn procedure (stage II) and the Fontan procedure (stage III). Stage I is performed in the first few days after birth. Stage II is performed at 3-6 months of age, and Stage III at 2-3 years of age.

The main goal of this strategy is to optimize the systemic circulation and to spare the heart as much as possible by creating a univentricular heart and a circulation in series in which the heart no longer has to maintain both circulations, but only serves as a pumping chamber for the systemic circulation. The procedures are all performed on cardiopulmonary bypass. The most essential points of the different steps of surgery will be described in the following illustrations.



Method: The illustrations were made to be used by surgeons, other specialists or students. Instead of showing them in an anatomical view, they are orientated, as seen by the surgeon in the operating room in order to address the surgeon immediately and to allow the reader to visualize the surgical field as the surgeon sees it. Therefore, I had to adjust to this view as well, in order to be able to understand the procedure and distinct the different structures (which was already harder as we dealt with an abnormal morphology and often partly reconstructed hearts). Despite that, I adapted to this new situation rather quickly, which made it natural for me to make the illustrations in this orientation.

The objective was to give a clear and detailed view on the surgical area. There are different techniques to make a suitable illustration. For instance watercolour or pencil. Instead, I chose to build up the illustrations in Adobe Photoshop in order to work as efficiently as possible. Often, small changes are made throughout the different steps of surgery. This Photoshop technique allows me to make easy adjustments without having to redraw the whole illustration every time again. Though this technique was new to me, I am happy with the time and effort I invested into it to bring it to this level. I built up the illustration in layers, starting with a green under painting, similar to the traditional watercolour technique, to save the overall uniformity.

Although, it would be proper to use a more graphic style for these kind of illustrations, I decided to make them more realistic. As the illustrations are made to be used by surgeons, they need to be more specialized than for patients for instance. The use of colours and shading was effective for the representation of the variance of tissues and textures, as a lot of different structures were involved, for example: instruments, fat, vessels, grafts... In addition, this technique gives me the ability to give a clear view on the surgical field, showing just as much information as needed.

The illustrations are based on sketches I drew upon direct observation, operative photographs and extensive consultations with the specialists. During the procedures, I did not always have a clear view because this surgical area in a neonate is very small and due to the constant beating heart. Therefore close communication with the surgeon was essential in order to convey not only the basic elements of the procedures, but also their most subtle details.

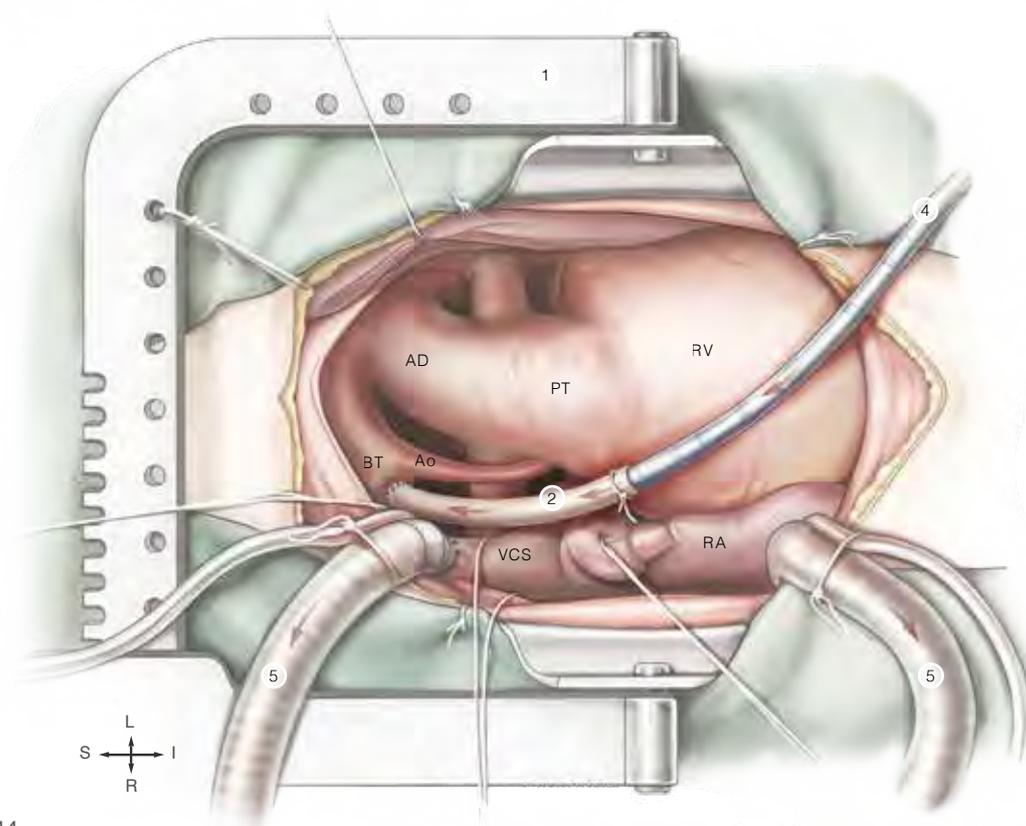


Figure 44
NORWOOD

After a sternotomy, the neonate is placed on cardiopulmonary bypass. The right atrial appendage is fixed with a stay suture to clear the surgical area. A graft (2) is connected to the brachiocephalic trunk and serves as a guide for the arterial cannula of the bypass (4).

CARDIOPULMONARY BYPASS

The cardiopulmonary bypass or 'heart-lung machine' is a machine, temporarily taking over the function of the heart and lungs during surgery. It is also considered as an extracorporeal circulation. It mechanically circulates the blood in the body, bypassing the heart and lungs, so the surgeon can perform the surgery having a clear and bloodless view on the surgical field. Two venous cannulas (5), one inserted into the superior caval vein, and the other via the right atrium into the inferior caval vein, drain the blood to the machine. The arterial cannula (4) supplies the oxygenated blood back into the body.

CIRCULATORY ARREST

Deep hypothermic circulatory arrest is performed during some major heart surgeries, which require surgery on the aortic arch, like the Norwood Stage I. It temporarily suspends blood flow under a very cold body temperature, causing the heart to stop beating, while preserving the organ function. The duration of circulatory arrest can go up to an hour. Nevertheless, it involves risks in an unstable neonate.

Figure 45
NORWOOD

First, an atrial septectomy is performed, through an opening in the right atrium. The interatrial connection is enlarged by removing a part of the atrial wall. The dashed lines indicate the incisions that will be made to complete further connections.

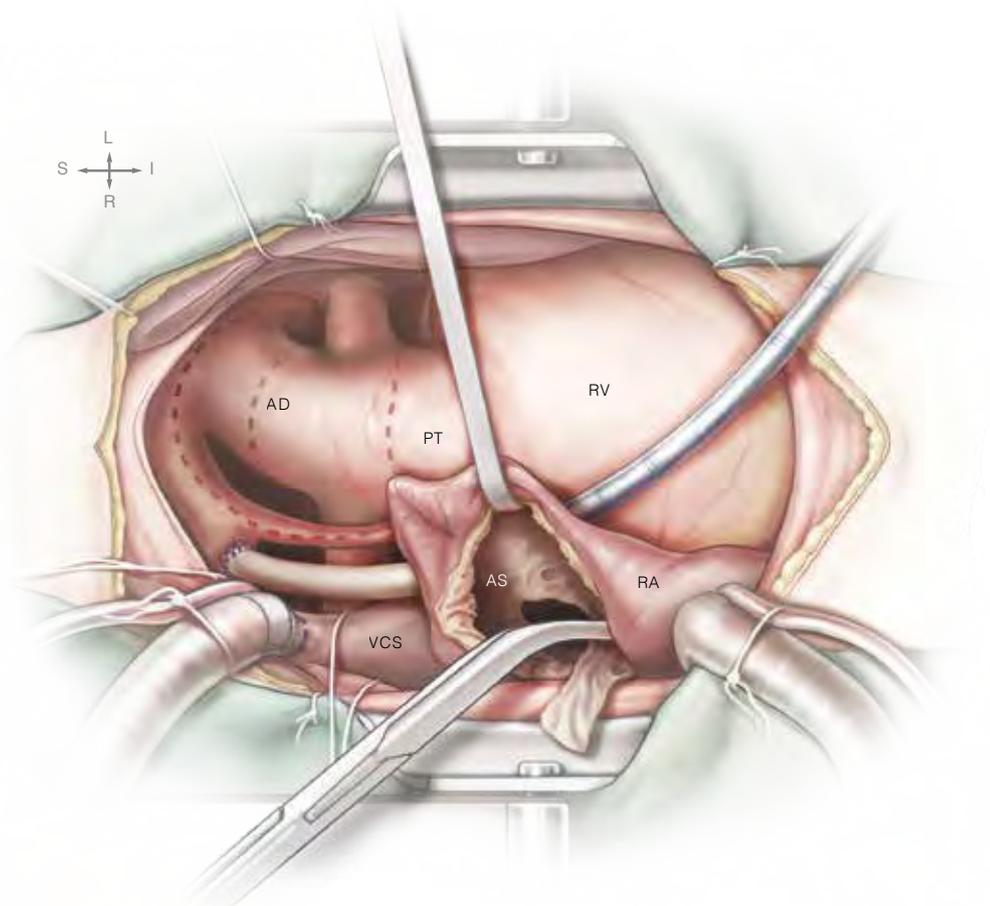


Figure 46
NORWOOD

The arterial duct is ligated close to the aorta and right before the trifurcation, and cut through in the middle. The pulmonary trunk is divided before the trifurcation, to disconnect the right and left pulmonary artery from the heart. The pulmonary artery is closed with continuous suture. The aorta is opened with a long incision as indicated in the previous illustration (aortotomy).

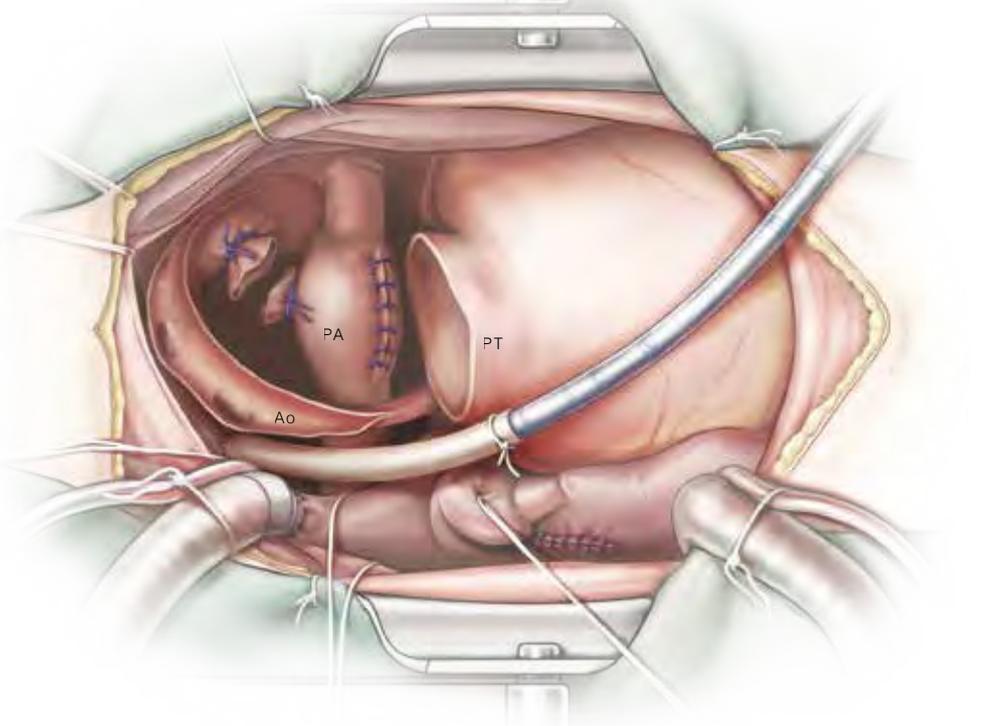


Figure 47
NORWOOD

A ³collar (3) is used to connect the small aorta to the large pulmonary trunk to create a bigger outflow vessel on the single working ventricle. The collar is first attached to the edges of the aortotomy. Then, the pulmonary trunk is attached to the collar, starting with the posterior part.

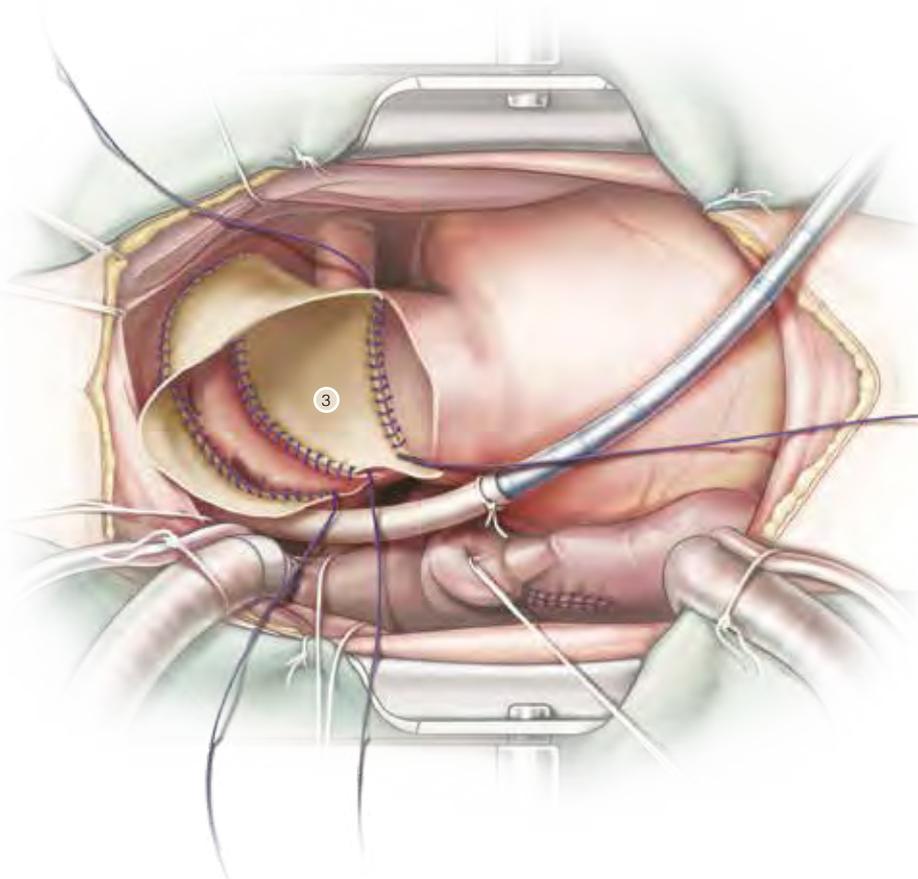
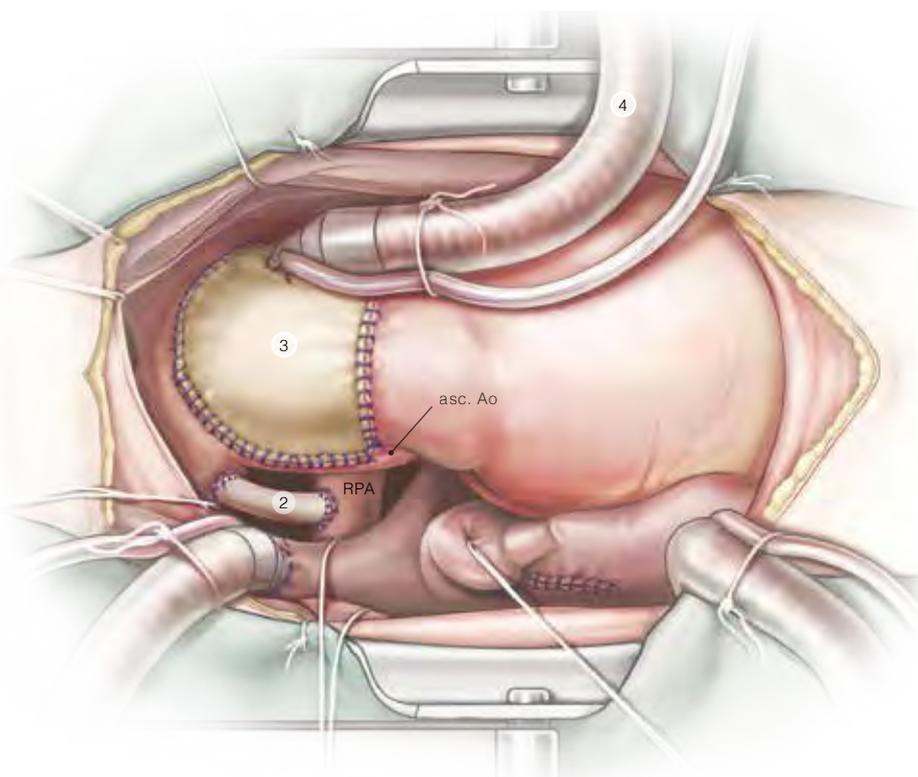


Figure 48
NORWOOD

After completing the anastomosis, a new arterial cannula (4) is inserted into the new common trunk. The cannula on the brachiocephalic trunk is removed and the graft (2) is connected to the right pulmonary artery. This systemic shunt serves as a conduit to supply blood to the lungs. The small intact part of the ascending aorta serves as a common coronary artery.



STAGE II

Figure 49
BIDIRECTIONAL GLENN

The gore-tex graft is removed. The superior caval vein is clamped close to the right atrium and more distally, not past the cannula of the cardio-pulmonary bypass. The dashed line indicates the incision that will be made to complete further connections.

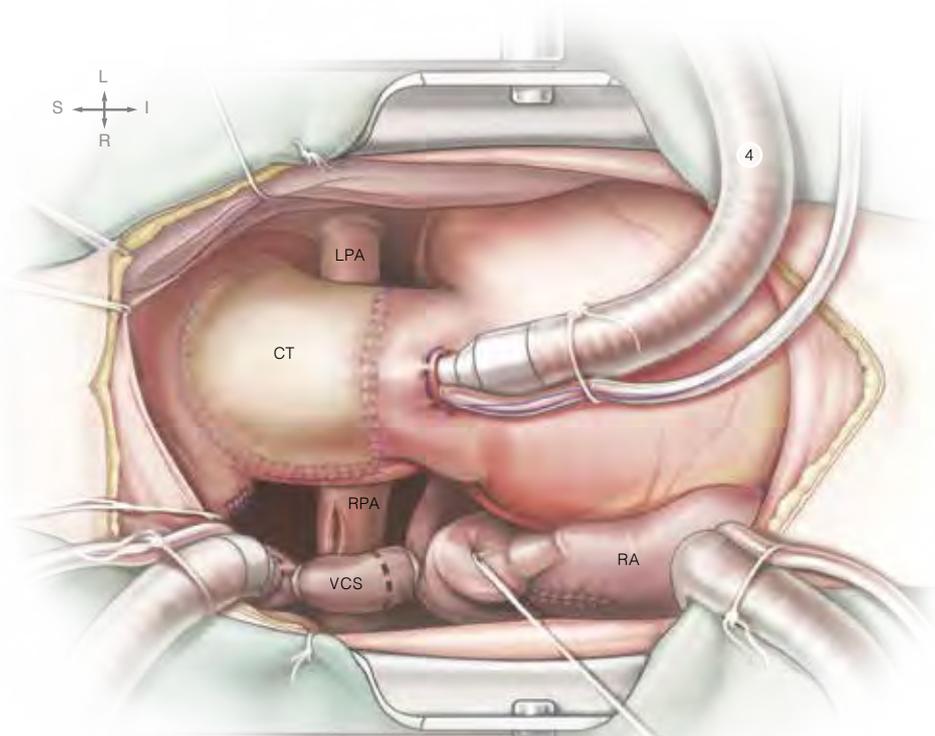
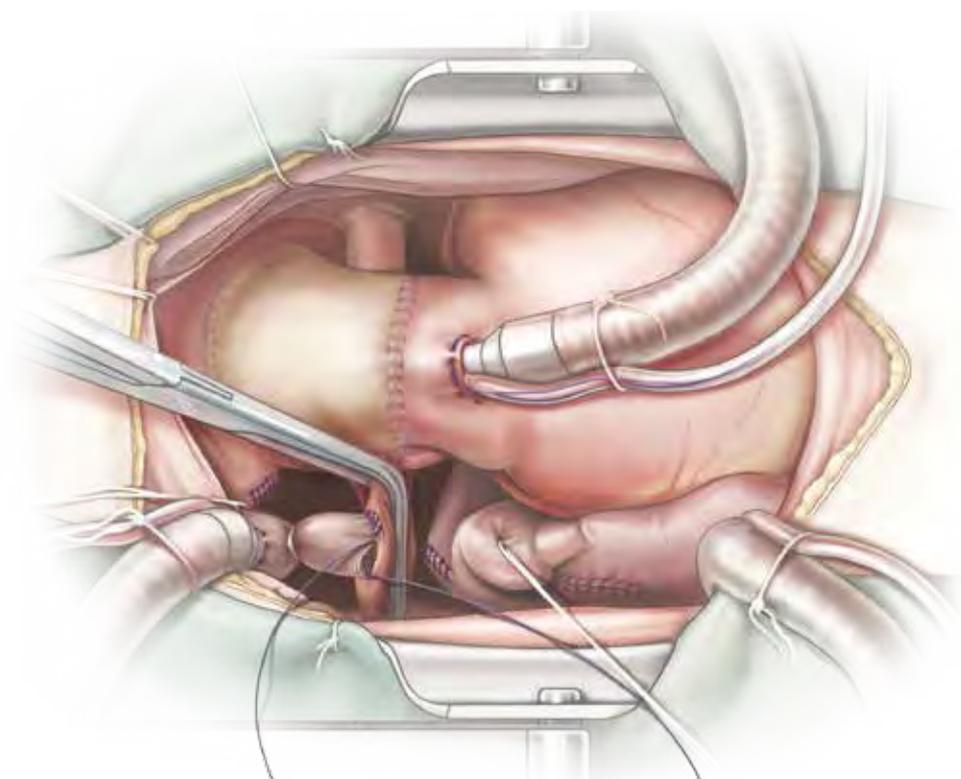


Figure 50
BIDIRECTIONAL GLENN

The superior caval vein is disconnected from the heart and the orifice at the right atrium is closed. A cavo-pulmonary shunt is made by connecting the superior caval vein directly to the right pulmonary artery. The desaturated caval blood, returning from the upper part of the body, will now directly pass on to the lungs.



STAGE III

Figure 51
FONTAN

The inferior caval vein is clamped at its orifice close to the right atrium, and pulled up, so the surgeon has a better view on these structures. The dashed line indicates the incisions that will be made.

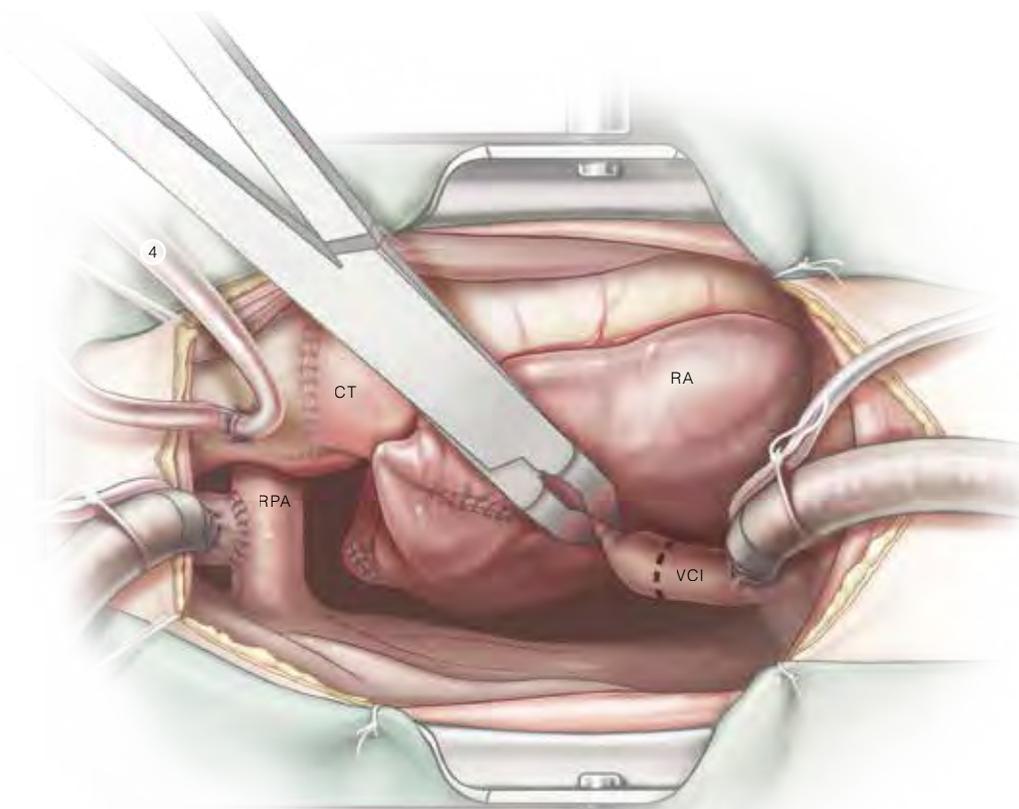


Figure 52
FONTAN

The inferior caval vein is cut through and the orifice at the right atrium is closed with continuous suture. A graft (2) is connected to the inferior caval vein with continuous suture.

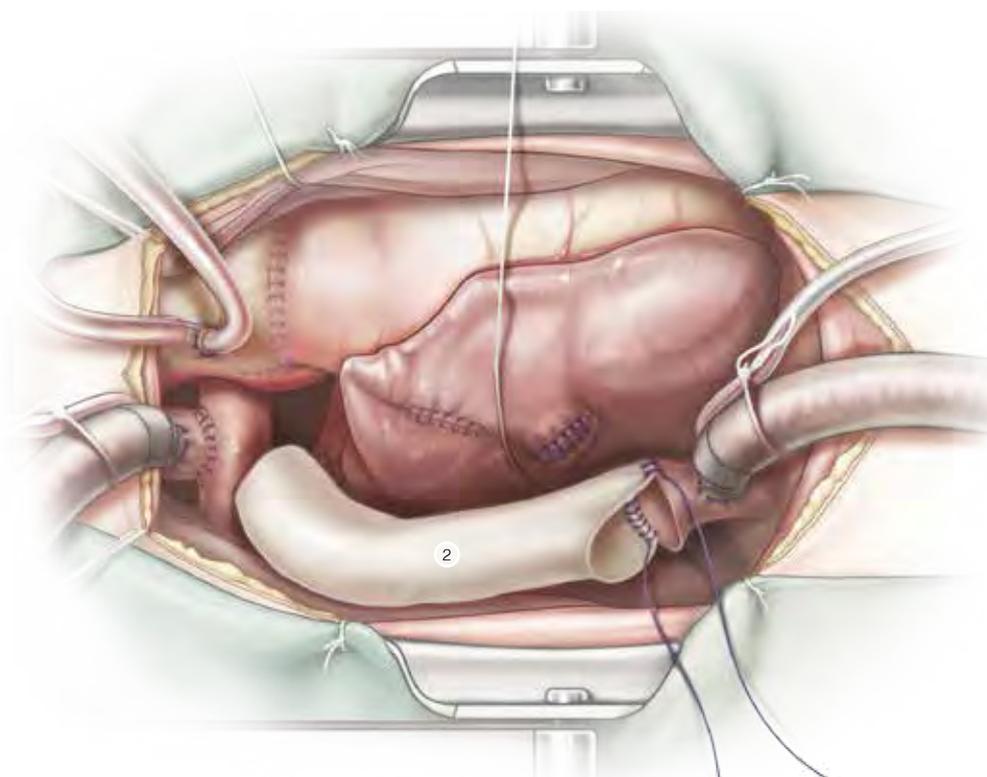


Figure 53
FONTAN

The anastomosis is completed by connecting the graft to the right pulmonary artery, close to the common trunk. This graft serves as a passage for the blood to bypass the right ventricle and go directly to the lungs.

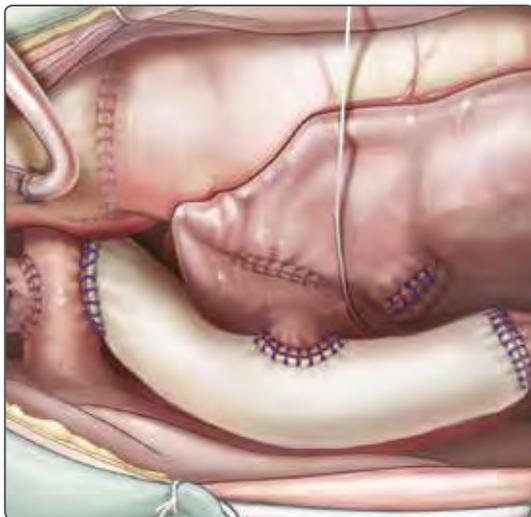
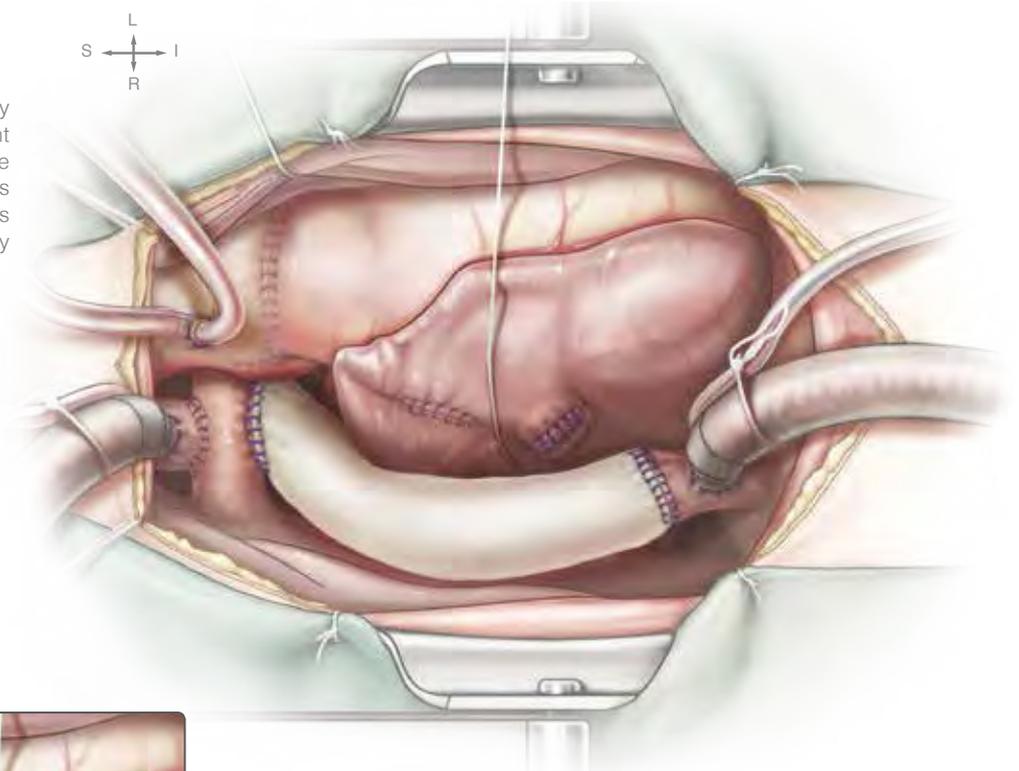


Figure 54
FONTAN

To avoid complications related to high systemic venous pressure, sometimes the surgeon makes a fenestration between the right atrium and the graft. When the pressure in the veins is high, a small amount of blood can escape through the fenestration into the right atrium and relieve the pressure.



Method: During the time frame of this Master Thesis, there was no suitable candidate patient for a Norwood stage I. Despite that, I needed to make illustrations of the procedure. After studying this cardiac malformation for a while and attending a hybrid procedure in addition, I was able to visualize the view on the heart as it would be exposed during the Norwood procedure. With the help of professional literature of the specialists, I could eventually reconstruct the different steps of surgery. First, we discussed the rough sketches I made. After some corrections, I worked out more elaborated illustrations in Adobe Photoshop.

Preparatory to the visualization of the Glenn stage II and Fontan stage III, I had the possibility to attend both procedures. The Glenn procedure was performed on a child with another type of cardiac malformation, which had not undergone a Norwood stage I. However, the steps of surgery were the same, so I projected this on the last illustration of the Norwood stage I. The use of different layer groups in Adobe Photoshop, enabled me to easily make adjustments after discussing the illustrations with the specialists. The Fontan procedure, eventually, was performed on a typical HLHS case. During this procedure I was able to check the illustrations of the Norwood and Glenn procedure, as the reconstruction made during the previous stages was clearly visible.

The sutures are coloured blue/purple, as this was a request of the surgeon. This makes sense because the sutures do have this specific colour, but moreover, it works very well in contrast to the colours of the other structures. To emphasize them in the illustrations, I decided to give the other sutures (stay sutures for example) a lighter colour, so they don't draw too much attention.

As the illustrations describe three stages of reconstruction, performed at different ages, the sutures of the first surgery would not be visible anymore during the last one. Despite that, I decided to keep them visible without being too dominant, by changing the transparency. Thereby, the reference to the previous illustrations is maintained.

FONTAN CIRCULATION

In a Fontan circulation, the systemic and pulmonary circulations are connected in series. All the blood, returning from the body, will bypass the heart and directly pass on to the lungs. The right ventricle volume overload is now reduced. There is no longer admixture of systemic and pulmonary venous blood. Only if a fenestration is made to relieve the systemic venous pressure, saturation is still lower than normal. The fenestration can be closed later during cardiac catheterization.

OUTCOME

Although the results of the surgical procedures have been improved over the years, the long-term outcome of the Fontan circulation remains uncertain and complications are common. Complications are often related to increase of the systemic venous pressure and chronic low cardiac output, and may lead to death. In case of ventricular failure, heart transplantation may be the only option left.

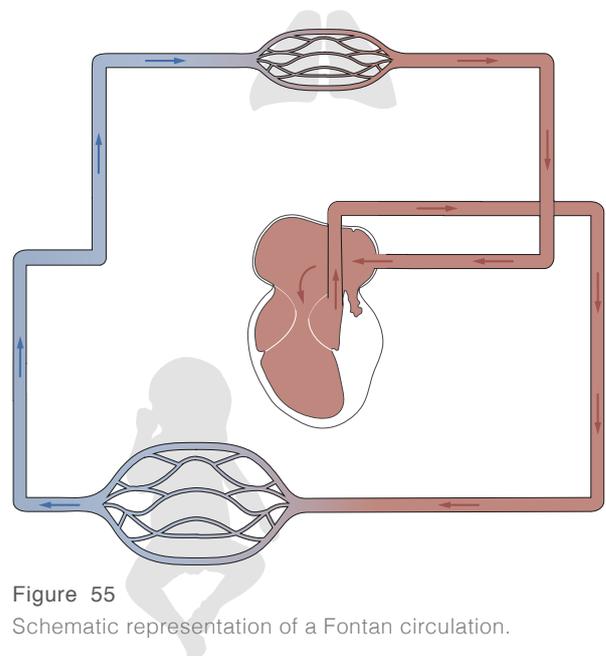


Figure 55
Schematic representation of a Fontan circulation.

3.2 HYBRID APPROACH

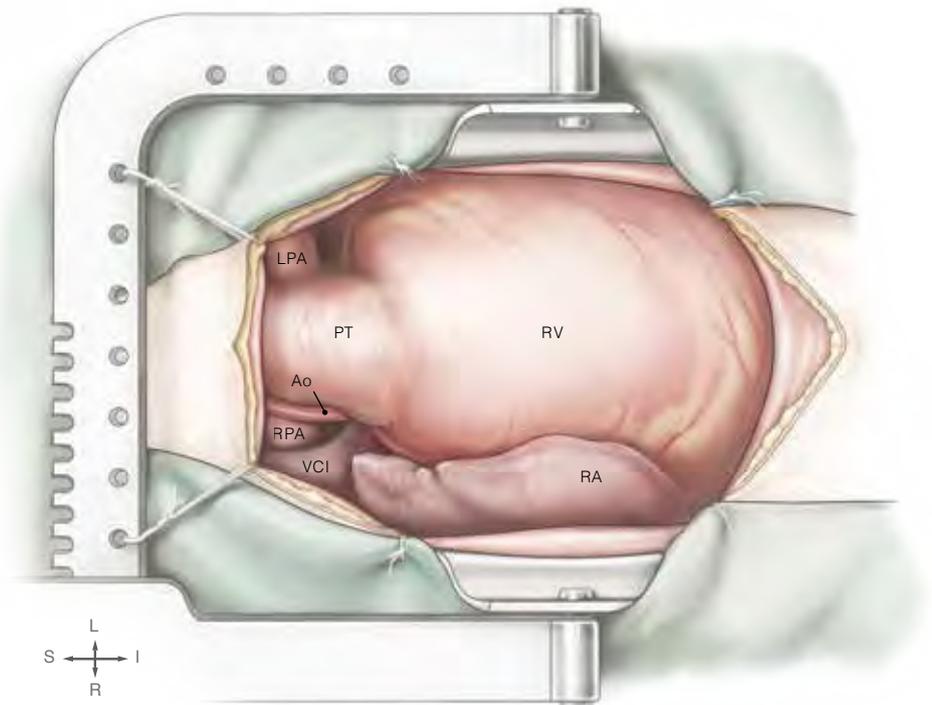
The hybrid approach has been developed as an alternative treatment for the management of HLHS. The initial intervention consists of three small and minimally invasive procedures: bilateral pulmonary artery banding, ductal stenting and a Rashkind balloon septostomy. These procedures are performed partly in the operating room and partly in the catheterization laboratory. Performing this hybrid procedure instead of a traditional Norwood procedure has the major advantage that the risk of major heart surgery can be shifted to an older age. It also creates more time for well consideration, allowing the option to proceed to a univentricular or biventricular repair as required.

BILATERAL PULMONARY ARTERY BANDING

The right ventricle overload results in an overload and dilation of the pulmonary arteries, causing a progressive increase of pulmonary blood flow and decrease of systemic blood flow. The primary objective of performing pulmonary artery banding is to improve the systemic pressure and ventricular function, and to reduce this excessive pulmonary blood flow and protect the pulmonary arteries from severe dilation and irreversible hypertension.

Figure 56
BILATERAL PULMONARY ARTERY BANDING

Bilateral pulmonary artery banding is performed through a median sternotomy approach, reaching a good exposition of the main pulmonary trunk and a good control of the pulmonary branches.



Method: As this hybrid procedure was the first procedure taking place during the time frame of this Master thesis, these were the first illustrations to make. Although this technique was new to me, I was able to bring it to this level eventually, after lots of trial and error. Pleased with the result, I continued making illustrations of the other procedures, which eventually took place at the end of my project. During this working process I noticed I got faster and it had been a great learning experience. However, after bringing all the illustrations together, I was aware that there was a huge difference between the last illustrations of the Fontan procedure and the illustrations of the hybrid procedure. During the process I discovered the benefits of combining graphic elements with a more realistic style. Outlines, for instance, give sharpness to an illustration and bring elements more towards the front. This was very useful to create a more three-dimensional image. I used an outline for structures such as: instruments, the surgical cloth, skin and fat, the pericard... To match the illustrations of the hybrid procedure with the illustrations of the other surgeries, I applied these outlines and changed colours where needed.



Figure 57
BILATERAL PULMONARY
ARTERY BANDING

During this surgical procedure, restriction of the pulmonary arteries is obtained by tightening (gore-tex) bands around the vessels.

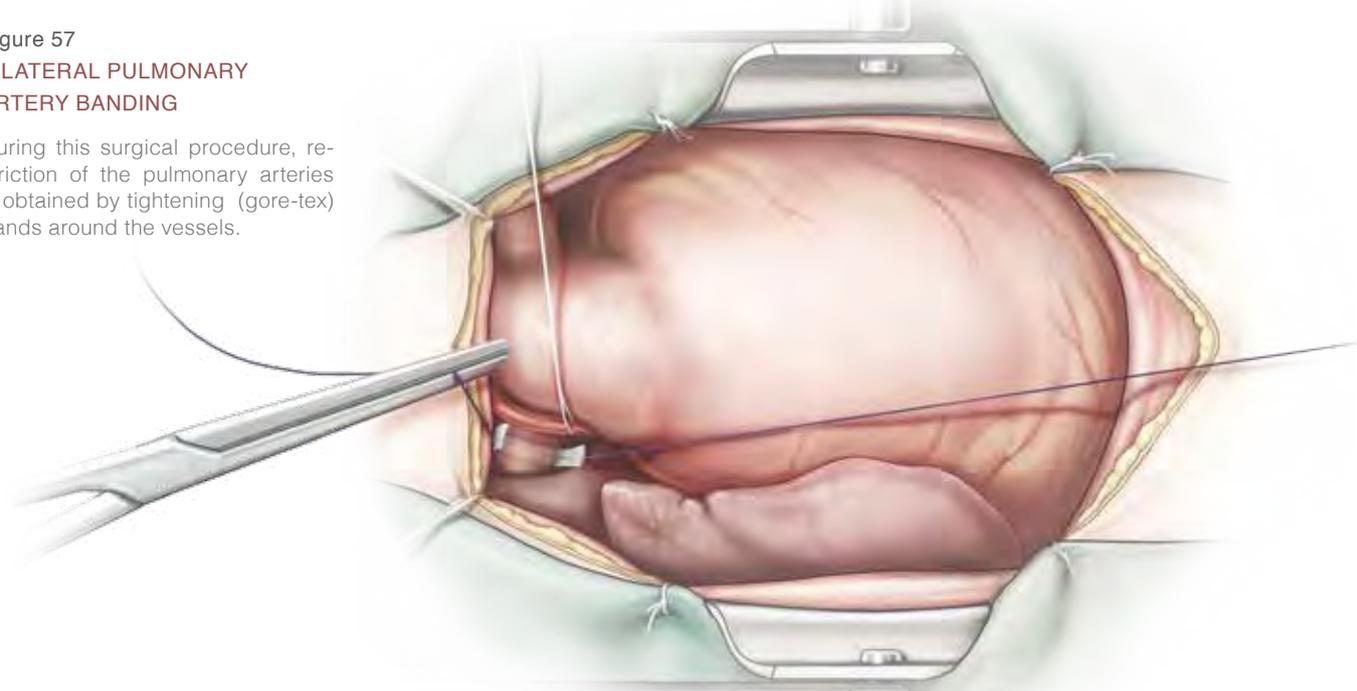
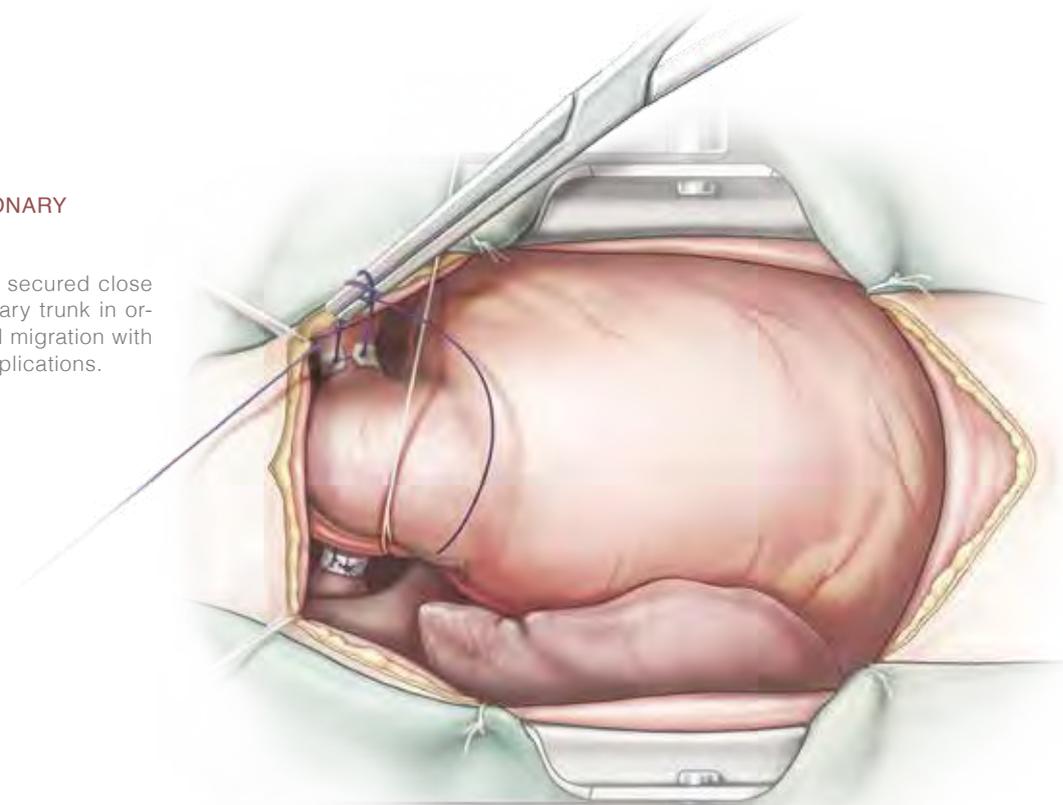


Figure 58
BILATERAL PULMONARY
ARTERY BANDING

The bands must be secured close to the main pulmonary trunk in order to prevent distal migration with its consequent complications.



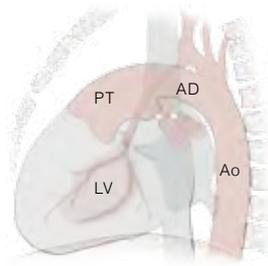
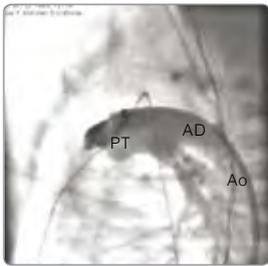


Figure 59

Left lateral chest X-ray during catheterisation exposing the pulmonary arteries and arterial duct.

After a contrast injection, a series of X-ray images is made to assist the cardiologist in locating the catheter and implanting the ductal stent in the correct position.

DUCTAL STENTING

This is the part of the procedure where the cardiologist takes over, starting with a stent implantation of the arterial duct. It will prevent the duct from closing and avoid obstruction of blood flow, which is important, as the neonatal circulation in HLHS depends on this connection for blood supply to the system. Right after birth, until surgical intervention is possible, it can be accomplished by using intravenous prostaglandin therapy to maintain duct patency.

The placement of a stent is a minimally invasive procedure. It can be performed either in the operating room during pulmonary artery banding, or later in the catheterization laboratory. Ductal stenting is usually performed under general anaesthesia. If the stent is implanted in the operating room during pulmonary artery banding, the most common access is through the pulmonary trunk.

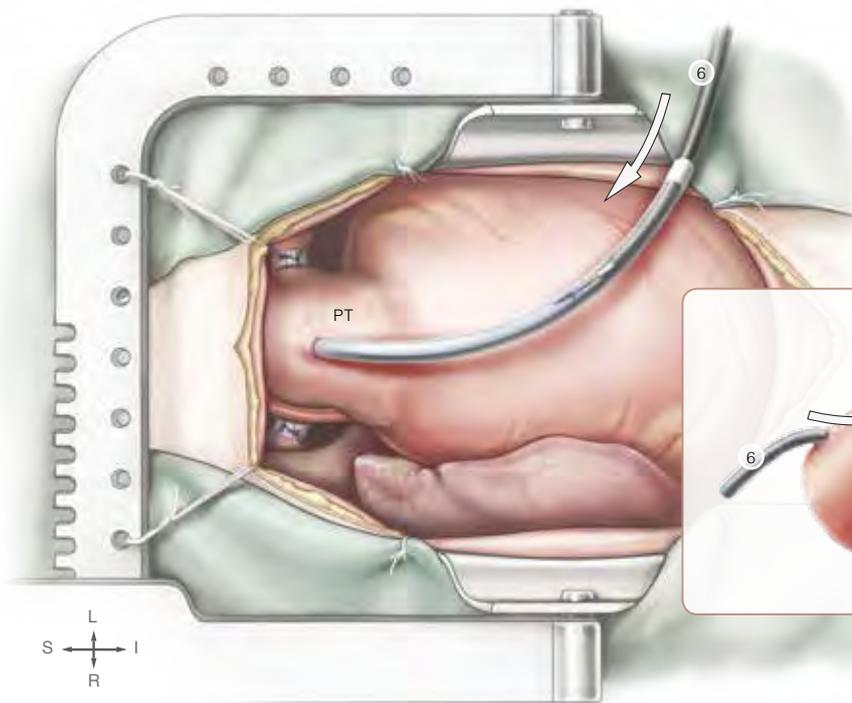


Figure 61
DUCTAL STENTING

The stent catheter (6) is inserted directly into the pulmonary trunk through a guiding catheter.

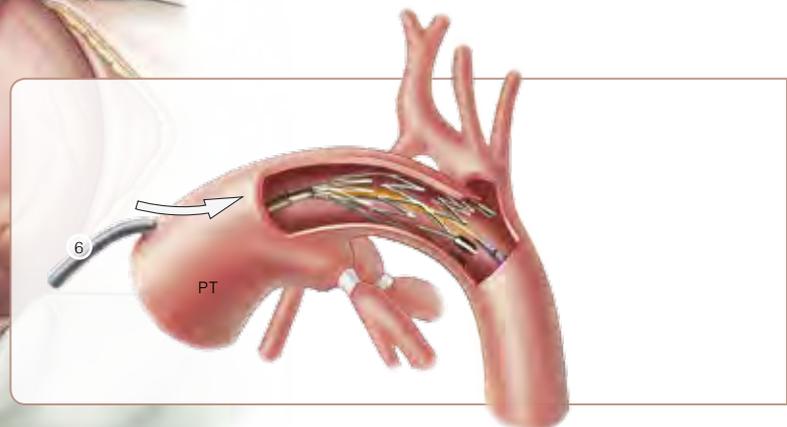


Figure 60

Left lateral view: placement of the ductal stent with access via the pulmonary trunk.

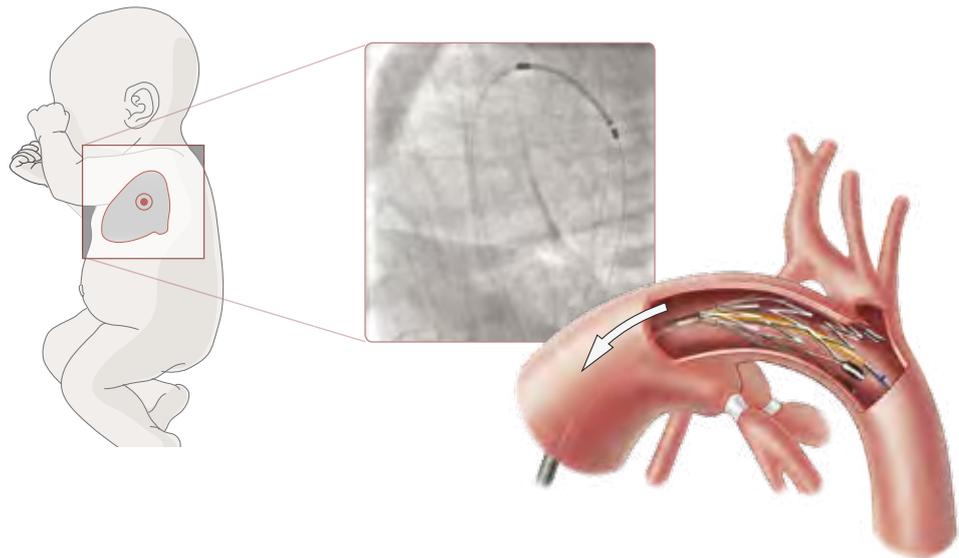


Figure 62
Left lateral view: after the catheter is positioned in the arterial duct (X-ray), the capsule is pulled back and the stent expands (Illustration).

If performed later in the catheterisation lab, femoral venous access is most common. However depending on the condition of the patient, retrograde arterial access may occasionally be used, although this is not preferable as arterial damage should be avoided.



Method: Although the paediatric and congenital cardiologists are expertised in this field, the radiological images made during cardiac catheterization may not always be clear to other specialists or doctors. Even though, they are very important in understanding the procedure and they give a lot of information about the heart defect as well. Therefore, I decided to immerse myself into the science of radiological imaging in order to gain more understanding of the subject. During cardiac catheterization a series of X-ray images were made, all providing information about different aspects. The exact size of the aorta and arch with its branches could be outlined with a contrast injection, while other images were showing only the movement of the balloon catheter (see next page) or the expanding stent. As an illustrator, I had the ability to combine the most important aspects of the different images to make an understandable illustration of the procedure. Again, communication with the specialists was essential. I decided to maintain the digital way of working for the same reasons as mentioned before.

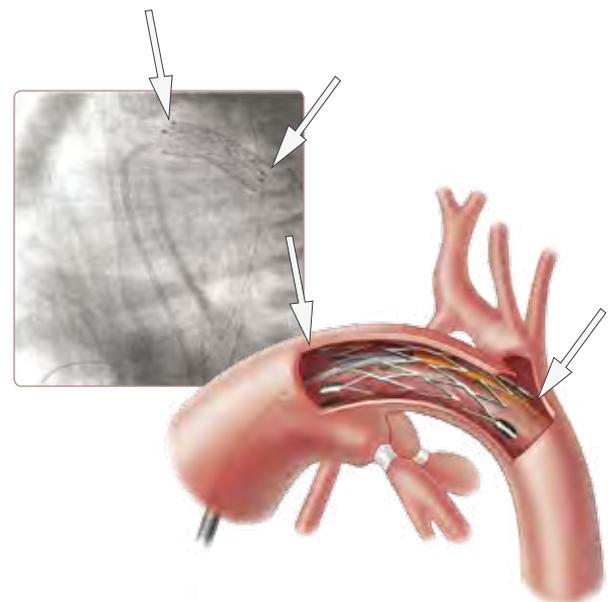
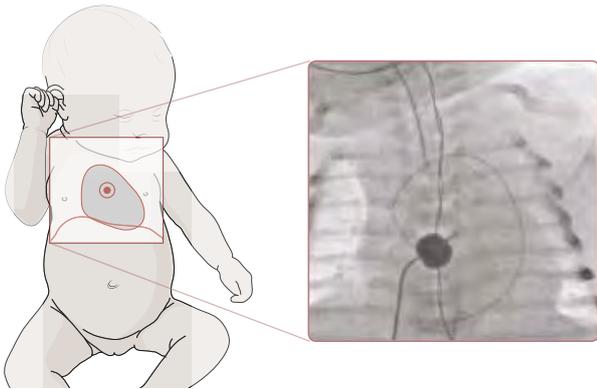


Figure 63
Left lateral view: the stent is implanted in the correct position, covering from just before the trifurcation of the pulmonary trunk till the insertion of the aortic arch.



RASHKIND SEPTOSTOMY

If necessary, a Rashkind balloon septostomy is performed in the catheterization laboratory. The main objective of this procedure is to enlarge the interatrial connection if it is restricted or partly blocked by the herniated valve of the oval foramen. This interatrial connection is important as it allows the saturated blood, returning from the lungs, to shunt into the right atrium, so it can be pumped out through the right ventricular outflow tract to supply blood to the systemic circulation via the arterial duct.

Access is achieved via the femoral vein. The balloon catheter is advanced from the right into the left atrium. Once the oval foramen is crossed and the catheter is positioned into the left atrium, the balloon is inflated and pulled with force through the oval foramen into the right atrium.

Figure 64

The balloon catheter (7) is advanced from the right into the left atrium. Once the oval foramen is crossed and the catheter is positioned into the left atrium, the balloon is inflated.

Figure 65

The balloon catheter (7) is pulled with force through the oval foramen into the right atrium in order to enlarge the interatrial connection.

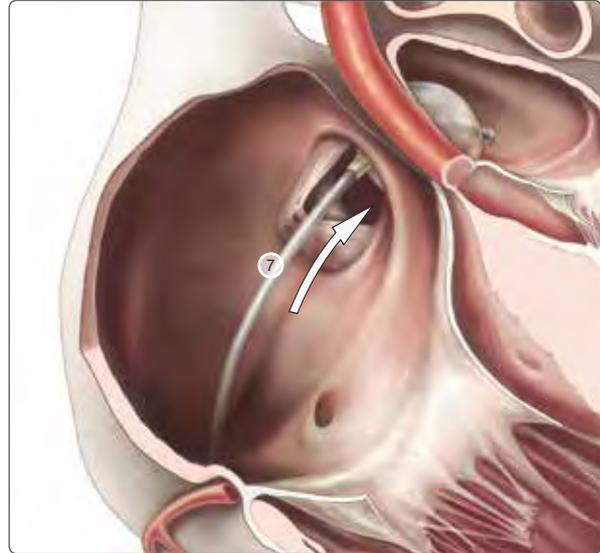


Figure 64

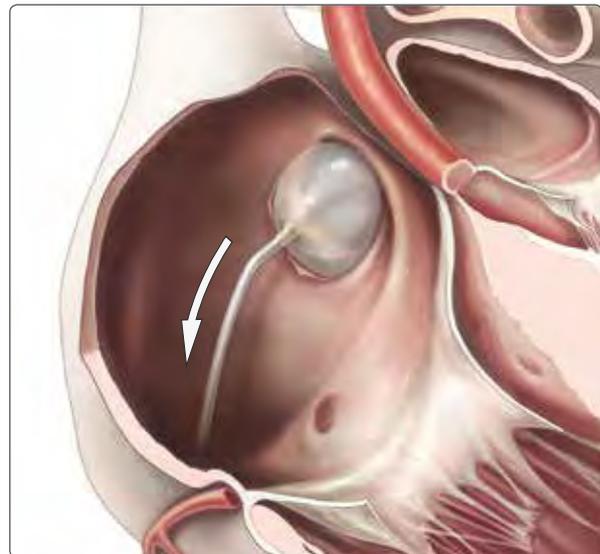


Figure 65

3.3 DECISION-MAKING

The management of HLHS remains a challenging and difficult problem. The outcome is strongly influenced by the severity of the left heart abnormalities. Specialists recently adopted the new hybrid approach as an alternative treatment strategy for HLHS, (and additionally) attempting to manage a larger number of patients with a biventricular repair.

The presence of severe ventricle hypoplasia and aortic atresia may demand a Norwood operation with a Fontan-type repair, as a biventricular repair in this case usually fails. However, in high-risk and unstable neonates, when major surgery is not an option at that stage, the hybrid approach can act as a bridge to univentricular repair. It allows adequate time for well consideration and the heart to improve, so major surgery can be shifted to a later stage. High-risk HLHS patients, such as patients with low body weight, aortic atresia, a restrictive atrial septum or poor ventricular function, are indicated as candidates for a hybrid palliation.

Also patients refusing to receive blood for religious reasons, such as Jehovah Witnesses, should undergo a hybrid procedure instead of a traditional Norwood stage I on cardiopulmonary bypass, because the latter procedure requires the admission of supplementary blood. This group of Jehovah Witnesses is an important subgroup as they were the initial reason for adopting this new approach.

Furthermore, previous complications have to be taken into account when deciding whether to proceed to a traditional Norwood stage I or a hybrid palliation.

Another indication for a hybrid palliation is a borderline left heart with growth potential of the left heart. In these patients, performing a hybrid instead of a traditional Norwood may allow the left ventricle to grow over time and reach an adequate size and function eventually to support a biventricular circulation. It also proved over time that in infants with HLHS, born to diabetic mothers, improvement of the left ventricular function could be expected.

The only exclusion criterion for a hybrid is echocardiographic evidence of restricted flow into the retrograde transverse aorta from the ductus arteriosus, due to coarctation.

In patients with aortic atresia, systemic circulation is dependent on retrograde bloodflow through the aortic isthmus. If there is any sign of obstruction or coarctation, the placement of a ductal stent may result in acute obstruction of the isthmus, leading to very low saturation. These patients will undergo a traditional Norwood procedure, unless bloodflow is secured by additional retrograde stenting of the coarctation.

Information regarding the treatment options and their associated risks and benefits is carefully presented to the parents. Although the treating specialists provide a recommendation for treatment and obtain informed consent, they have the obligation to respect the parent's final decision.

4

FOLLOW-UP AFTER HYBRID

4.1 UNIVENTRICULAR REPAIR

In case there is no growth potential and biventricular repair is not possible, the patient is scheduled for a comprehensive 'stage II' surgery at 4-6 months of age.

COMPREHENSIVE STAGE II AND FONTAN COMPLETION

This procedure is performed on cardiopulmonary bypass with aortic crossclamping. Circulatory arrest is not needed because of the growth of the transverse aortic arch. During this major heart surgery, the ductal stent and pulmonary artery bands are removed, the aortic arch and pulmonary artery are repaired if necessary. Further on, it consists of all steps of a traditional Norwood and bidirectional Glenn procedure (usually, a septectomy is not needed as a Rashkind septostomy is performed earlier during the hybrid procedure).

To complete the reconstruction, a Fontan procedure is performed as a stage III. The cavo-pulmonary connection is completed and there is no longer admixture of systemic and pulmonary venous blood.

OUTCOME

Although the comprehensive stage II is a long operation, the postoperative results are better than those of traditional Norwood stage I, because the patients have a lower risk profile (due to improvement of the ventricular function and more stabilized circulation) when going into the operation.

There may also be an impact on the neurologic development of the patient, as most patients with HLHS have a fragile central nervous system. Placing them as a neonate on cardiopulmonary bypass and

circulatory arrest, can lead to neurological injury. When the hybrid stage I is performed instead of a traditional Norwood stage I, cardiopulmonary bypass and circulatory arrest are shifted to later in life when the infants' brain has grown and developed more, with its benefits on the long-term neurologic outcomes.

However, the growth of the pulmonary arteries after debanding can be problematic. And nice pulmonary arteries are essential for a good Fontan.

4.2 BIVENTRICULAR REPAIR

In some cases of a borderline left heart, the placement of a ductal stent and pulmonary artery bands may allow the left ventricle to grow over time, due to the additional blood flow into the left ventricle. The left-sided structures of the heart may show sufficient growth potential to reach a normal size and function during the postnatal development, and to allow a biventricular circulation.



Figure 66
Echocardiography A (four-chamber view) on a neonate with a borderline left ventricle.

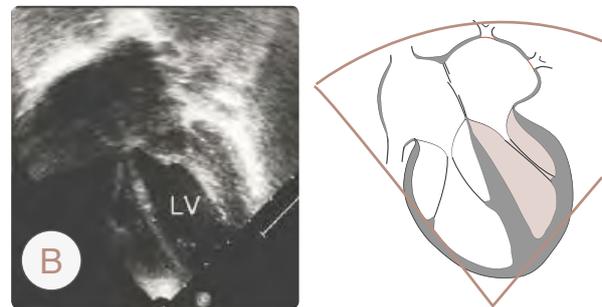


Figure 67
Echocardiography B (four-chamber view) months after treatment the left ventricle cavity has significantly increased.

ROLE AS ILLUSTRATOR

Besides the complexity of the subject, the biggest challenge for me as an illustrator in this project was definitely the question how to deal with the lack of good reference material. In anticipation of a suitable candidate patient, I tried to gain as much information as possible out of professional literature, articles and interviews with the specialists.

As I had to make a start somewhere, I decided to begin with some drawings of a normal neonatal heart and further, an HLHS case. Based on the gathered information and images out of books and from the internet, I tried to 'reconstruct' these illustrations. However, after a first meeting with the specialists, I realized there were a lot of small mistakes, although I looked very closely in the books. For example, an opened aortic valve looked completely different than how it was presented in the books. After the specialists showed me a real valve, I noticed the difference and I was able to make a correct illustration.

Also the arch of the aorta of a neonate with HLHS seemed to be rather different. As I started from the normal anatomy, I didn't regard the abnormal function and flow in case of this cardiac malformation. The retrograde flow caused a completely different morphology. This made me realize how important it is to fully understand and know every aspect of this cardiac malformation, in order to be able to create correct illustrations. And furthermore, drawing from first hand observation is by far more preferable than interpreting from third party reprints.

Only after having a chance to observe a particular procedure, I would actually have the ability to make an accurate interpretation of the morphology of HLHS as it is exposed.

I was privileged to have the possibility to attend a hybrid procedure early in this project. The patient suffered from a very severe form of hypoplastic left heart syndrome with aortic atresia and mitral valve stenosis. Although this case did demand a univentricular repair, a hybrid procedure was performed as a bridge to univentricular repair, in order to gain more time for the patient to improve and stabilize. However, pulmonary artery banding is performed through a medium sternotomy so the heart was not fully exposed.

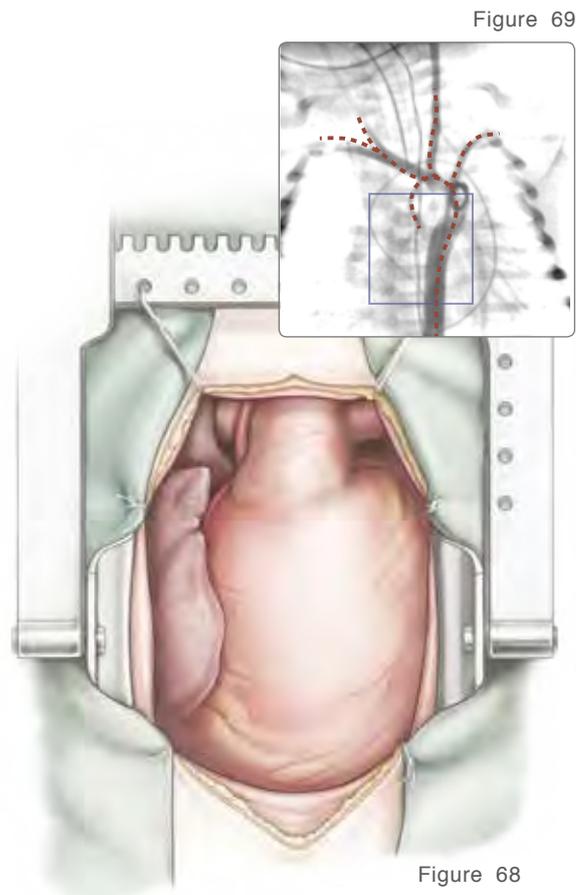


Figure 68-69

During pulmonary artery banding the heart was partly exposed (fig. 68). The aorta and branches could be outlined during cardiac catheterization (fig. 69).

I observed and registered the parts of the heart shown during surgery. The aortic arch, with the subclavian and common arteries and the arterial duct were not exposed during surgery but could be outlined on X-rays later during cardiac catheterization. I projected the part of the heart I registered during surgery on the X-ray in order to be able to reconstruct the missing parts. This image I used as a template to make a rough sketch in Adobe Photoshop of the complete heart. In the end, I decided to only use the outlines and make a clear drawing in Adobe Illustrator. This illustration matched the descriptions of prof. dr. Gewillig and prof. dr. Rega.

For the visualization of the other cases of the spectrum, the only elements I could truly rely on were the knowledge and experience of the cardiologist and the cardiac surgeon.

And this was the goal: to succeed in visualizing their ideas and knowledge to make a complete, clear and accurate overview of the spectrum of HLHS. For that, of course, close cooperation and good communication with the specialists was essential.

Figure 70

A rough sketch in Adobe Photoshop based on the X-ray and the part of the heart seen during surgery.

Figure 71

The aorta and its branches could be outlined during cardiac catheterization. This was useful in reconstructing the aortic arch correctly.

CONCLUSION

This project has been a challenge in all aspects and it has increased my skills at so many levels. Although I really appreciate the splendiddness of a detailed traditional pencil or watercolour illustration, in this case it was not effective, as I only had little information to rely on. It forced me to look for other solutions. It provided the possibility to practice my digital skills. Although this felt out of habit at first, I worked out an effective system to make schematic and detailed illustrations that are easy adjustable, emphasising only the essence. These illustrations did perfectly fit the needs of the target group. During this process I learned that a simple line drawing in Adobe Illustrator can be even more usefull in some cases than an elaborated pencil drawing. Moreover, using this digital technique, I was able to finish this large number of illustrations within the time frame of this Master Thesis.

I also learned to deal with radiological images as an important source of reference, and to find solutions for the vizualisation of this kind of complex procedures.



Figure 70

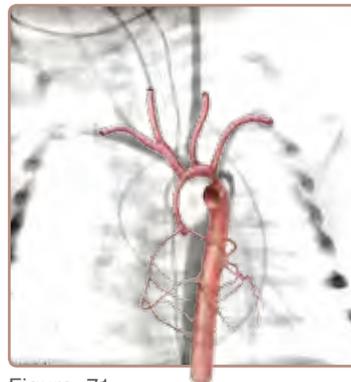


Figure 71

When it comes to ethics, I learned the importance of staying focussed on the subject, as we dealt in this case with very young patients in critical conditions. Furthermore, the importance of close cooperation and communication with the specialists really improved my professional skills. Their critical and motivating attitude worked positively stimulating. This resulted in a constructive interaction.

To conclude, I think we succeeded together in creating a good overview of the spectrum of HLHS and the different treatment strategies.

SAMENVATTING (NL)

Hypoplastisch linker hart syndroom of HLHS is een aangeboren hartafwijking, met slechts enkele gevallen per jaar in België. Bij kinderen met HLHS is er een variabele hypoplasie of onderontwikkeling van het linker gedeelte van het hart, waardoor er onvoldoende zuurstofrijk bloed naar het lichaam gepompt kan worden. Voor de geboorte is circulatie nog mogelijk door de aanwezigheid van de ductus arteriosus en het foramen ovale. Vlak na de geboorte echter sluiten deze twee foetale connecties en komt het kind ernstig in gevaar.

Tot voor kort was de Norwood procedure de enige optie voor kinderen met HLHS. Dit is een reconstructie bestaande uit drie operaties met als doel het univentriculaire hart zo optimaal mogelijk te gebruiken en zo min mogelijk te belasten. Deze openhart operaties, waarbij gebruik wordt gemaakt van een hart-long machine, zijn echter niet zonder risico voor een onstabiele neonaat. Verder is de prognose van een univentriculaire circulatie ook niet zo optimaal. Daarom hebben specialisten recent een nieuwe hybride strategie ontwikkeld voor de behandeling van HLHS, met zeer goede uitkomsten. Deze procedure bestaat uit drie minimaal invasieve procedures, waarbij chirurgische en cardiologische technieken worden gecombineerd. Het grote voordeel is dat het gebruik van de hartlong machine vermeden wordt bij risico patiënten. Het doel van deze strategie is een stabiele circulatie te creëren, en daarbij de mogelijkheid te behouden om naar een univentriculaire of biventriculaire circulatie te gaan, naargelang de behoeften van de patiënt.

De complexiteit echter van de afwijking en de ernst van hypoplasie maken ieder geval uniek en telkens weer een uitdaging om te behandelen. Daarom hebben de specialisten in Leuven alle gevallen opgedeeld in drie groepen, volgens de ernst van hypoplasie. Deze verschillende vormen van de afwijking zijn systematisch beschreven en geïllustreerd in het tweede hoofdstuk. De illustraties van de nieuwe hybride procedure in het derde hoofdstuk dienen als aanvulling bij de artikels die recent verschenen zijn over deze nieuwe strategie. Aangezien van deze nieuwe strategie en van het spectrum van HLHS nog geen duidelijk overzicht was met goede illustraties, ligt hierop de focus in dit project.

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BIBLIOGRAPHY

Mark Galantowicz (MD), John P. Cheatham (MD), Alistair Phillips (MD), Clifford L. Cua (MD), Timothy M. Hoffman (MD), Sharon L. Hill (ACNP), and Roberta Rodeman (RN). (2008). *Hybrid approach for hypoplastic left heart syndrome: Intermediate results after the learning curve*. by The Society of Thoracic Surgeons

Stephen C. Brown, Derize Boshoff, Benedicte Eyskens, Marc Gewillig. (2009). *Hybrid approach as a bridge to biventricular repair in a neonate with critical aortic stenosis and borderline left ventricle*. by the European Association for Cardio-Thoracic Surgery

Marc Gewillig. (2005). *Congenital heart disease: The Fontan circulation*

C. Gittenberger-de Groot, *Het links hypoplastisch hart als aangeboren afwijking*

John W. Kirklin, Brian G. Barratt-Boyes. (1993). *Cardiac Surgery* (vol. 2), second edition. New York, US: Churchill Livingstone.

Frank H. Netter (MD). (1971). *The CIBA collection of medical illustrations: heart* (vol. 5). New York, US: CIBA.

Wallace A. McAlpine. (1975). *Heart and coronary arteries: an anatomical atlas for clinical diagnosis, radiological investigation, and surgical treatment*. Berlin, Heidelberg, New York: Springer-Verlag.

Benson R. Wilcox, Andrew C. Cook, Robert H. Anderson. (2005). *Surgical Anatomy of the Heart*, 3rd edition. UK: Cambridge University Press.

T. B. Johnston, C.B.E. (MD), J. Whillis (MD), (MS). *Gray's Anatomy*, 13th edition. London, New York, Toronto: Longmans, Green and co.

Michael Schünke, Erik Schulte, Udo Shumacher, Markus Voll, Karl Wesker. (2010). *Prometheus: inwendige organen*, 2nd edition. Houten: Bohn Stafleu van Longhum.