# Gresham College Main logo

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**The Heart: Introduction to the Series**

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## [Slide 2 Thematic Slide + Title for Series]

Provost, faculty and staff of Gresham, Ladies and Gentlemen, friends, colleagues and my family.

I am deeply honoured to have been appointed the 37th Gresham Professor of Physic and to be speaking to you today. It is truly humbling to take ones place in such an historic chain, to recognise the achievements of my predecessors and to have the opportunity to meet, and work with, the fascinating people who have become my new colleagues at Gresham College.

I hope you will have found a small box on your seat when you arrived. It contains props you will need later. So look after them and try not to tip them up.

It is particularly poignant to be able to deliver my lectures here at the wonderful Museum of London. As a family, my wife Lesley and my son Becan have supported the 1983 part of the Timeline of London in memory of Toby, Becan’s brother, who died in 2009.

## [Slide 3 Photos of Toby]

I would like to dedicate this lecture series to his memory.

## [Slide 4 The Heart; an Introduction]

This, the first in my series of lectures, is designed to introduce to the heart and particularly those aspects related to those problems which occur during development and which result in congenital heart defects. But first I want to spend a few moments explaining why I have chosen the overall theme and consider its relevance, if any.

## [Slide 5 Movie of Heart Surgeon at work…fading into background]

I have spent more than 35 years looking after children with mostly congenital abnormalities of their heart and lungs. This specialty in which I work is tiny, and it is reasonable to ask ‘what possible relevance might it have to a general audience?’ Particularly when you set the scale of the problem it faces in context.

## [Slide 6 Causes of Death in children]

Congenital disease makes up only a small proportion of the causes of death in children, and heart defects make up and even smaller % age of those, with an incidence of <1% of live births. By far the most common causes of death in childhood are related to infections, and many of those can be attributed to infection, most of those are intimately related to under nutrition.

## [Slide 7 improvement in mortality of 20y]

In fact, childhood mortality has fallen significantly over the last 20 years in association with the Millennium Goals projects. Almost half that reduction has come from better management of pneumonia, diarrhea and measles, not to improvement in the management of congenital heart defects. Perhaps I should be talking to you about sanitation, public health and good governance if I want to make a difference.

Sadly, most of this mortality is concentrated in Asia and particularly sub-Saharan Africa.

## [Slide 8, Gresham background]

So why bother you with congenital heart disease? Well there are personal and, I hope additional rational reasons to be interested. The personal reason is that surgical treatment for congenital heart defects is contained largely within the span of my life. I was born in 1951, and before the 1950’s there were very few reparative treatments available for congenital heart defects. Modern heart surgery was born roughly when I was, and development was rapid.

The specialty that comprises the management of congenital heart disease has features which can act as a lens through which to consider the scientific, ethical, financial, political and organisational changes in healthcare in the late 20th and early 21st centuries. The specialty has lived through, and benefitted from, truly disruptive technological changes, such as the development of the heart-lung machine and computing. Materials science has radically altered our field, and imaging is now spectacular. The specialty has lived in the public eye. Its members have boasted about their achievements, and the media have loved them, sometimes treating them as celebrities.

It was the first medical discipline to be ‘transparent’ about its outcomes.

It has benefitted and suffered from such extensive public scrutiny, been a battleground for politicians and seen significant changes in culture, including exemplifying teamwork at its best. Its rate of progress and technical challenges have continued to attract pioneers and encourage collaboration with other disciplines, and the simple motivation of working with children and their families has added to a relentless drive for excellence.

There has been remarkable international co-operation in development and education and few would argue against its success.

## [Slide 9 Mortality for Surgery for CHD]

Here is a graph that summarizes the success of the subject in the course of my lifetime. Prior to 1950, mortality was approaching 100%, now in centres like Great Ormond Street, it is down to less than 2%. I was attracted to the subject by the drama of headlines such as ‘hole in the heart baby lives’, it is a testimony to the advances we have made that the headlines now are more likely to be ‘hole in the heart baby dies’.

I will discuss many of these topics in more detail in the course of my lecture series, but I want to start today by considering many of the anatomic, physiologic, physical and philosophical reasons which make the heart such a fascinating and evocative subject.

## [Slide 10 natural curiosity]

Humans just love to know about their insides, as French commented in 1978. And with good reason; our insides perform some pretty miraculous work.

## [Slide 11 Gilgamesh]

Yet, as early as 2600 BC, Gilgamesh, King of Uruk in Mesopotamia, witnessing the death of his friend (in battle of course…little changes in that area) is said to have uttered these words “*I touched his heart, but it does not beat at all*”. This is thought to be the first reference linking the heart to its life sustaining function.

## [Slide 12 heart animation]

This remarkable organ starts beating within a relatively few days of conception…and just keeps going. It beats over ***100,000 times a day, 40 million times a year and 3 billion times in an average lifespan***. And it works as the engine room of a delivery and collection system involving over ***97, 000 km*** of vessels. A remarkably reliable logistics network. Later in this lecture I will outline how this amazing organ develops, and what the consequences of failure of that development might be. But it is worth mentioning here that the heart usually still keeps on working, even when significant parts of it are malformed. Astonishing.

## [Slide 13 soul etc]

But it is not just the form and function of the heart which fascinate. The heart has held a special place in human culture for thousands of years.

It has acquired figurative importance as the seat of the soul, emotion, strength and indeed of humanity itself. These philosophic concepts of both the heart and its role have persisted for millennia and are reflected in many phrases current parlance and literature.

## [slide 14 of book of dead, balancing feather]

The ancient Egyptians held the heart in high regard. The Book of the Dead, from the 16th C BC Papyrus of Ani, reveals a little of their view. Here it is shown being weighed after death against a feather, representing **truth or divine order**. The heart was seen as important not only in **the judgment of truth**, but also as **a measure of self-worth** in the afterlife. The Egyptians treated the heart with great reverence during embalming, replacing it in the body or preserving it in a jar….unlike the brain, which was pulled out piecemeal via the nose with a hook, as shown here:-

## [slide 15 brain hooks etc]

best not to try this at home, but it is good to know the old Egyptian ways live on!

## [slide 16 slide of xin]

In ancient Chinese philosophy, the heart was considered the centre of human cognition. That **Xin**, a term which occurs frequently in Confucian writings and those of his followers, and which is usually translated as ‘**heart-mind’**, reflects this.

## [Slide 17 Sanskrit/Hindi]

The heart is stated in the Rigveda to be crucial to making moral judgements, and even Empedocles

## [Slide 18 Empedocles]

who was the force behind the Sicilian medical schools in the 5th C BC believed the heart to be the most important organ in the body, and ‘the seat of the soul’

## [Slide 19 Philolaus]

Philolaus, probably from Crotone in Calabria, at roughly the same time, got closer to the function of the brain, but not the heart, and certainly hadn’t cracked the purpose of the umbilicus

## [Slide 20, the Bible with credits]

The Bible too is littered with cardiac references. In the Old Testament, the heart was the locus of intellectual, moral and psychological **life**, a theme which persisted into the New Testament.

## [slide 21 al-Ghazzali]

In Islamic tradition too, the heart was perceived, as Al-Ghazzali (1058-1111) says **‘the monarch of the entire body’.**

## [slide 22 the heart is a pump]

Al-Ghazzali was closest to the truth. The heart might be a metaphor for the spirit, but in reality it is just a pump, albeit one at the centre of our circulation. Getting to grips with what the heart does and how it does it, rather that what it might signify has indeed taken millennia.

## [slide 23 astonishment replaced by reasoning and interfering]

As van Tellingen put it ‘astonishment was replaced by reasoning and interfering’.

Classical era theories of the way the circulation worked were complex, and (largely) fundamentally wrong. Preeminent amongst these theories, and certainly the longest lasting, were those of Galen.

## [slide 24 Galen’s circulation]

Galen was a Greek physician of the 2nd century AD. He was an intellectual force to be reckoned with, and his ideas went largely unchallenged for almost 1500 years, indeed they were still actively taught in the 16th century.

Sir Henry Dale and Sir Thomas Lewis made a film in 1928 to celebrate the 300th anniversary of Harvey’s publication of de mortu cordis. Sir Henry updated this for the Royal College of Physicians in 1957, and I want to show you a wonderful clip from that movie.

The movie demonstrates Galens concepts of life giving forces, concoction and the expulsion of ‘fuliginous vapours’ via the lungs. And his complete failure to grasp the function of the valves, the unidirectional nature of blood flow and the danger of mixing air with blood in the heart.

## [slide 25 Thomas Gresham]

The ideas of these classical scholars certainly evolved with time, but it was the circulation as we know it was not appropriately understood until around the time of Sir Thomas Gresham.

## [slide 26 Leonardo da Vinci]

Leonardo’s wonderful anatomical drawings clearly demonstrate the important cardiac structures, including the valves, the significance of which eluded Galen and his followers.

## [slide 27 Versalius, Columbus and Harvey]

But the circulation was finally resolved by Harvey and his immediate predecessors, Versalius and Columbus, in Padova.

## [slide 28 Harvey’s observations]

Harvey understood the important unidirectional nature of the flow of blood in the veins back to the heart by this classic observation of the function of venous valves , and understood the relationship between the pumping of the heart and the peripheral pulse, it is thought because he observed the function of fire-pumps in the street.

## [slide 29 Thomas Gresham]

Much of this work coincided with the start of the Gresham chairs around the late 16thCentury. Well might Thomas look rather doubting. My predecessors in this post lived through some very exciting times in London, but did not always make choices in the interest of public health and, it seems, may not have got to grips with some of the seminal science of the day.

## [slide 30 Physic Timeline 1]

Matthew Gwinne was the first Professor of Physic, appointed in 1597. The years between his appointment and that of his successor, Peter Mounsell, were, to say the least, eventful.

After the plague closed in theatres in London in the early 1590’s, the Globe opened in Southwark in 1599. In 1600, the East India Club was founded and in 1603, Elizabeth 1 died. In 1604, James VI of Scotland became James 1 of England, and in the context of these lectures, published a remarkably prescient document…his **Counterblaste to Tobacco**, in which he said……”***a custom loathsome to the eye, hateful to the nose, harmful to the brain, dangerous to the lungs, and in the black stinking fume thereof nearest resembling the horrible stygian pit that is bottomless”.*** It is a shame that it took us another 400 years to clear the air.

1605 brought the Gunpowder Plot, and 1606 not only the hanging drawing and quartering of the plotters, but also the first debate (failed) in the Act of Union in parliament. 1611 saw the publication of the King James bible, and in 1615 Thomas Winston became the 3rd Gresham Professor of Physic.

## [slide 31 Physic Timeline 2]

In 1620, the first Physic Professor, Matthew Gwinne, became a commissioner for tobacco, a role described later as ‘unscrupulous Government officials controlling the licensed traffic in tobacco’. Obviously, I cannot know if that applied to Gwinne, but it certainly looks like a wrong turning. In 1628, Harvey’s great work, “de Mutu Cordis”, was published, which according to Wikipaedia, unfortunately had little impact on Thomas Winston, and did not stop him becoming closely involved with tobacco companies.

You can perhaps see why I am nervous that what I say I these lectures will prove seriously flawed in due course!

The work of Harvey really set the scene for modern cardiovascular medicine, yet the ability accurately to diagnose and surgically to manage congenital abnormalities of the heart is relatively recent. But before we discuss that in more detail, let us look at the miracle which happens every time a baby develops.

## [slide 32 Pregnancy]

This short film, which I have accelerated a bit, shows you how quick is the rate of development of the baby in the womb. You can see the heart beating in the very early weeks after conception, driving the distribution system for oxygen, nutrients and the removal of waste products during growth, returning them via the placenta to the mother. The astonishing rate of development soon becomes a recognizable infant, and leads at 40 weeks to the delivery of the fully formed baby. I am constantly amazed that this incredibly complex and highly integrated process is so remarkably reliable, and so rarely goes wrong.

The next lecture in this series will consider how we fix things that have gone wrong with the heart, but I need to introduce you to concepts of development, morphology and physiology Which Have Formed the background for that work and which continue to fascinate those of us in the field. Lets first see what how the developed heart works.

## [slide 33 Animation of beating heart]

Your heart is a muscular pump divided into right and left sides. Bigger than your fist, it sits slightly left of center in your chest. It pumps between 5-8 liters of blood around every minute, and as we said about 100,00 times per day.

Oxygen-poor blood, "blue blood," the oxygen having been removed by the body, returns to the right atrium heart. The blood passes from right atrium across a valve to keep the flow going in one direction to the right ventricle and through another valve into the artery to the lungs, the pulmonary artery The lungs refresh the blood with a new supply of oxygen, making it turn red, and getting rid of CO2.

This oxygen-rich blood, "red blood," return to the left, composed of the left atrium and ventricle, and is pumped through the aorta to the body to supply tissues with oxygen.

When your heart contracts it is called systole and you can feel the effect of this in peripheral arteries as your pulse. The whole thing sis synchronized by an inbuilt electrical system which initiates and distributes contraction of the specialised heart muscle.

## [slide 34 basic diagram of the circulation]

That circulation of blood is shown here in a diagram which I have made to form the basis of how we might get to grips with congenital heart disease. Blue blood, with the oxygen used up by the body returns to the right atrium, passes through the tricuspid valve to the right ventricle and then out to the lungs via the pulmonary valve and arteries. From there, now red with oxygen added, it passes back to the left atrium via the pulmonary veins and from there across the mitral valve into the left ventricle and out through the aortic valve to the aorta and the rest of the body.

The resistance to the flow of blood through the lungs is much less than than it is in the rest of the body, so the right ventricle has to do less work to pump blood through the lungs and the left has to do more to pump it round the body. The pressures generated in the chambers of the heart are thus different as you can see on the slide. These pressure and resistance difference s are very important to understanding the physiology of what happens when development of the heart goes wrong.

How does complex and compact organ develop? Until 20 years or so ago, understanding of the development of the heart was based on observation, serial sections and reconstructions of human embryos, and to a large extent, speculation. Reconstruction is still important, but the use of genetic constructs in the mouse has made it possible to trace the fate of cells expressing genes of interest, and thus map, the movement and changes in those cells. This may be described as an evidence-based molecular view of development. Let’s have a look at how the heart does develop, based on this science.

Here I want to acknowledge first the help of Dr Andrew Cook of UCL who is an expert in the development of the heart, and Jacob Read and Matt King who are, as you will see, wonderful animators. They have worked together, selflessly, to develop this short film both for us here at Gresham, but also for future students of the field to help them understand what goes on.

## [slide 35 animation of the heart]

This starts with fertilization at day zero! The sperm and the egg join as two nuclei in a single cell. That cell divides into two, four and then eight by day 3. Multiple cell divisions occur until it is about .5mm across and between days 4 and 5 it condenses to create a disc of cells (the embryonic disc) with fluid above it. This embryonic structure implants itself in the lining of the uterus.

Looking at it from above, embryo next organises itself into left and right halves, helped by rotating cilia or hairs in a small node, the primitive node, which appears at the base of the embryo. A midline structure also appears, on either side of which form two ‘heart forming fields of cells’. The primary heart field is shown in red, and the secondary heart field in blue.

These fields then migrate forwards towards the head end and become the cardiac crescent. We are now at about day 15, and the whole embryo is less than 2mm.

These heart forming fields then expand and increase in size, and looking from the side, you can see them bulging more at the top, and being fused together at the top and the bottom. At these points of contact, the secondary heart fields in blue, donates cells to the primary heart fields.

This movement of cells, especially at the upper end, creates the outflow areas of the heart, and results in complex looping or bending of the heart with some ballooning of shape, resulting in the creation of the two ventricular chambers.

All animals have arteries arranged in segments initially, and these undergo regression and merger. At about 5 to 8 weeks the arrival of more cells from a part of the embryo called the cardiac neural crest catalyses the division of the single way out of the heart into separate aorta and pulmonary trunk. The embryo is now 4-5mm in length.

A second set of workmen arrive, this time from the top of the liver, these are cells from the pro-epicardial organ. These cells cover the surface of the heart and reorganize to form the coronary arteries and veins.

Another tube of cells emerges from the lungs and moves towards the back of the heart to create the pulmonary veins, allowing blood to flow back from the lungs to the atrium.

All this is happening at roughly the same time, from 5-8 weeks, and if we open the side of the right ventricle at this time, we can see that there is work going on inside too. Walls are being built between the two atria at the inlet to the heart and between the two ventricles in the main pumping part and the wiring is being installed. Three so called cushions associated with the areas where valves will develop also form, this all creates a 4-chambered heart with the left and right sides separated by walls, and the top and bottom separated by valves, and with a working electrical system.

Paradoxically, a new hole appears between the atrial chambers to allow blood from the placenta to flow across to the left side of the heart. This hole usually closes after birth.

## [slide 36 left and right]

We make all sorts of assumptions about left and right sides of the heart, but why is there left and right at all? Why not symmetry? Well, all sorts of cool things go on in the primitive node. There is a pumping mechanism moving cells from right to left in this example, driven by lots of rotating flagellae or cilia; tiny hairs. It is these that make the parts of the heart left and right and dictate their position. If these go wrong your heart can inverted, or you can have two left sides or two right sides. Way too complicated to explain in detail, but I love these little things.

## [slide 37 The scale of things]

It all looks very beautiful on the big screen, but in reality the very size of the heart is itself amazing. In your box you will find a walnut and a grain of rice. The newborn heart at 40 weeks gestation is almost always describes as ‘the size of a walnut’ rather like on the News, everything is the ‘size of Wales’. I could not afford to put a pound coin into your box, but as Andrew shows here, at 20 weeks the heart is no bigger than a pound coin, and at 14 weeks no bigger than a grain of rice. Yet t functions throughout this time, and goes on your 3 score years and 10.

You can keep the props, and the box. But remember if you watch the next lecture just how small everything is and imagine operating inside it.

The heart may have a simple job to do, but as you have seen from its development, the final arrangement of the heart within the body is both complex and elegant. It is a triumph of compact function and efficient packaging. Sadly, many things can go wrong during that development and result in abnormalities of the heart described as Congenital Heart defects.

Anatomic or more accurately morphologic descriptions tell you what the defect *looks like* and where it is, but not *what it does.* But they are necessary to communicate and compare. It was only when surgical approaches began to be developed, and particularly the development of open-heart surgery that there was accelerated interest in the description and classification of anomalies. Progress was most rapid as echocardiography developed.

Pathologists and anatomists had identified some heart defects over the course of history, and have long argued as academics do over how exactly to classify them. However, their contribution to understanding has been immense, and surgeons, cardiologists, radiologists and most importantly computers now speak the same language, even if as you will see our perceptions are different.

## [Slide 38, sequential segmental analysis]

To describe the defects, a systematic approach has to be taken and the concept of sequential segmental analysis was evolved, largely from work by Lev in Boston in the 1950s.. The heart is considered in 3 parts; the atria, the ventricles and the great vessels. In each, the component parts are described, their connections are noted and associated lesions which affect blood flow listed. This logical sequence drives the process of diagnosis as we shall see.

## [Slide 39, IPCCC]

There followed something like the Ryder Cup of cardiac morphology, with an American team led by Richard van Praagh and a European team led by Bob Anderson, who became both friends and foes! Van Praagh’s group took the view that the *relations* between the segments was of primary importance, but Anderson and colleagues took the view that the way the segments were *connected* was of greater importance. Those of us who lived through all this debate often felt we were in the middle of a Victorian argument on semantics; Atriums or atria, suprachristal or perimembraneous oulet; superior caval vein or a superior vena cava and so on? *Ad Infinitum!* What did it matter it was what you saw, so why all the fuss? Passions ran high, and international meetings were, to say the least, partisan.

It did matter of course. Medicine is about the transfer of information. If you can’t exchange that information and have agreed definitions of what is wrong with a patient, how can you realistically agree diagnoses or compare treatments or outcomes? A referral letter may simply describe the wrong thing and initiate the wrong course of action. It became even more important when echocardiography developed as more people than just a few experts had access to diagnostic tools and were able to exchange data more freely. The agreement between the US and Europe fostered in the 1990’s by the International Nomenclature Society, of which for my sins I was president at the time, has made unity of database possible, and allowed meaningful comparison of results.

## [Slide 40, list of diagnoses]

Thousands of individual things can be wrong with your heart, as this scrolling list of IPCCC codes demonstrates, and there is a lot to fix. Because the names are long and often problems are multiple, professionals often abbreviate them creating an excluding code, but one which allows us to work faster. For example transposition of the great arteries with perimembraneous outlet VSD and a patent ductus arteriosus becomes TGA, PM outlet VSD& PDA. This can be tough for families and patients, but it is a constant surprise to me how quickly most of them learn, if they have to.

Fortunately, the process of sequential segmental analysis allows us to simplify (a bit) into groups of problems, some of which I shall now outline. Some of these will be mentioned in later lectures, but there is no exam at the end, so it is not necessary to remember the detail, just the principle.

## [Slide 41, basic diagnoses]

Before I start that, I want to tell you how, until the late 1970’s we diagnosed these conditions. It was largely through detailed knowledge of pathology, combined with a great deal of clinical skill and a lot of inference. Patients were either blue (cyanosed) or pink depending and there was either not enough or too much blood going to the lungs. We were able to deduce diagnoses from the noise of the flow of blood in the heart (murmurs), as you can hear in these two examples, and further deduce things about the heart from its electrical activity on ECG. Over time more detailed anatomic diagnosis has supervened.

## [Slide 42, basic heart diagram]

Let us see what can go wrong, starting first with our basic diagram. Some of the most common congenital anomalies are abnormal or persistent communications between the chambers. If there is a communication between the two atria it is called and Atrial Septal Defect or ASD.

## [Slide 43, ASD]

A classic ‘hole in the heart’ and the first to be repaired.

When there is such a hole in the wall between the atria, blood will flow from the side with the high resistance (the red side in this diagram) to that with low resistance, creating a left to right shunt.

## [Slide 44, ASD L to R shunt]

Those right-sided chambers thus carry more blood and get a bit bigger, and the valve close a bit later…you can hear that late closure of the pulmonary valve in this clip.

## [Slide 45, ASD R to L shunt]

If, for any reason the resistance to flow of blood through the lungs is high, as in some specific diseases then the flow of blood across the hole will be reversed to Right to Left, leaving the patient with more blue blood in the left side of the circulation and thus blue extremities or lips, which is called cyanosis.

## [Slide 46 ultrasonography]

I said earlier that diagnoses were inferred from a variety of clues. All that change with ultrasonography, developed, again at the start of my life, by Hertz and Adler in Lund. Cardiac ultrasound emerged a decade later…albeit with massive machines.

## [Slide 47, better echo machines & Doppler]

The digital revolution has seen better and now smaller and smaller devices being used to demonstrate anatomy, and by the application of the Doppler principle, the velocity and direction of blood flow can be both quantified and visualized in glorious Technicolor!

## [Slide 48, ASD, echo]

This slide shows first a clip from our developmental animation. If something goes wrong with that process, holes may be present in a number of sites as shown here in this diagram. ECHO clearly shows such holes, painlessly and non invasively, and we can see flow, left to right in this case, and also reconstruct the ASD in 3D and in real time .

## [Slide 49, ASD, MRI]

In certain circumstance, more functional detail is required, and for this we use Magnetic resonance. Here is an example.

## [Slide 50, VSD]

Another relatively common communication between chambers is the ventricular septal defect or VSD. Here the hole is in the wall between the ventricles, and again there are significant variations in detailed location, but the net effect is the same. Blood flows from the high resistance side to the low resistance side, but across a bigger pressure gradient. This is a L to R shunt at ventricular level. Too much blood goes to the lungs, which become ‘wet’ and the child short of breath. Eventually the lung arteries become damaged by this excess flow and resistance gradually rises eventually, over years, reversing the flow from left to right to right to left, ending up making the child blue. By this stage the child is usually inoperable.

## [Slide 51, VSD, echo and MR]

Here some example of VSDs, again showing the developmental model, the sites of holes and the echo appearances. In both 2D - with the shunt flow from left to right, and in 3D. The next panel shows multiple VSDs in 3D, and in the last a complex VSD imaged by MR.

## [Slide 52, missing components]

## [Slide 53, absent RV]

The next significant group of anomalies includes those with missing or absent components. For example, the right ventricle may be missing creating a single ventricle or univentricular heart. In these circumstances the arteries beyond the ventricle are also often small and to survive blood must cross from L to R at atrial level, and get to the distal lung arteries via either a natural communication called the ductus arteriosus or a man made shunt from a branch of the aorta to the lung artery.

## [Slide 54, hypoplastic RV]

In fact ventricles are rarely completely absent. They are much more often very small or hypoplastic. The net effect is the same, with the same need for shunting of blood to survive.

## [Slide 55, hypoplastic RV echo]

Here is a good example of such a small ventricle, the shunting of blood it produces, and the inevitable cyanosis.

## [Slide 56, 57, absent LV]

We will see in later talks that one of the most controversial topics on our field over recent years has been the management of small or hypoplastic left ventricle. The LV can be completely absent with a left to right shunt at atrial level and again the need for a shunt a great arterial level to supply the body, but more commonly the left ventricle is just very small, as shown here

## [Slide 58, hypoplastic LV]

The need for shunting remains the same.

## [Slide 59, Echo and MR of HLHS]

Here you can see a small LV on echo, the shunting of blood and the same thing on MRI although you have to excuse it being back to front.

These patients also almost always have very small aortas and these need reconstructing after birth.

## [Slide 60, abnormal connections]

We have seen abnormal communications, absent or small parts, and now I want to show you some abnormal connections.

## [Slide 61 TAPVC partial] [Slide 62, TAPVC total]

The first of these is partial anomalous pulmonary venous connection, in which the veins coming out from the lung have joined up with the wrong atrium. Here just three, but we often see all the veins taking a devious and wrong route to the right atrium, ending with too much blood going to the lungs, and dependency on an atrial communication, or an emergency operation, for life.

## [Slide 63, 64, transposition of the great arteries]

The final example I want to show you is also one to which we will return in later lecture. That is transposition of the great arteries, in which the aorta and pulmonary artery come off the wrong ventricles…they are transposed. This results in a very dangerous circulation in which blue blood comes back from the body, only to be sent there again, and red blood comes back from the lungs, itself only to be sent back again, but already fully loaded with oxygen. The body does not get enough oxygen, the baby is blue and survival is only possible with an ASD, PDA or both.

## [Slide 65, perception]

it is all very well having nice, relatively simple diagrams like this to explain how things look and work out what needs to be done. Sadly we all have different perceptions and our understanding of reality is relative. I am sure many of you will have seen this, but it makes the point.

If I ask you what colour are the two squares labeled A and B, I am sure that most of you will say grey or white. In fact they are both the same colour, and our eyes are working out what to expect from surrounding squares and interference patterns at the line margins. But our eyes are wrong.

## [Slide 66, the cardiologist and the surgeon]

Here is an echo of a problem called atrio-ventricular septal defect…a combined hole in both the septa. It looks great on the echo, and you can clearly see the holes. It looks big too.

But in reality, the surgeons see something completely different, and within the walnut sized heart, with no margin for error. The more we can understand of the morphology preoperatively the better, and you will see in another talk how working as a team helps.

## [Slide 67, 3D stuff]

Also of great help has been the advent of 3D reconstruction, either virtual or by printing. Here is a 3D reconstruction of a heart in which one of the coronary arteries takes origin from the artery to the lungs rather than the aorta. It is clear to see and an operation can be planned quite easily.

In this montage we see the outlet from the right ventricle in several different patients who have had previous surgery and need a valve implanted. Not only can we see the variation in shape, but also dynamic change, and this combined with 3D printing allows us to design and rehearse procedures for the individual child.

## [Slide 68, counter, life]

We have only been able to fix these many problems in the era since I was born

## [Slide 69, Gresham time]

If the life of Gresham College represents 12 hours, that is just 1.5 hours of Gresham time.

## [Slide 70, next time]

Next time I am going to discuss how we have come to be able to fix these things.

Thank you for listening, and thank you to all the people who have helped me put this together, and tolerated my absence or demands.

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