Surgery for Congenital Heart Defects; science or art?

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"There is nothing more difficult to take in hand, more perilous to conduct, nor uncertain in its success, than to take the lead in the introduction of a new order of things. For the innovator has for enemies all of those who have done well under the old, and lukewarm defenders in all of those who may do well under the new."

Niccoló Macchiavelli (1469–1527)

In my first Gresham lecture, *[The Heart; an Introduction (http://www.gresham.ac.uk/lectures-and-events/the-heart-an-introduction)]*, I outlined how special the heart is to people, how our views of its form and function have changed over time and how it develops. I described a few of the more common abnormalities which can occur during that development. I also pointed out that the ability to repair most congenital heart defects itself only evolved after the introduction of open heart surgery in 1952, almost paralleling the duration of my life.

In this lecture, I want to mix a bit of history with some personal observations, and reflect on the significant disruptive technological advances that have occurred during those 60 odd years. I am not going to repeat the *detailed* histories of the subject which are available elsewhere(1-7), rather I want to pick out some highlights and consider the characteristics of the early pioneers of cardiac surgery, and how later cardiac surgeons acquired their, sometimes gruesome, reputations for autocracy and arrogance. It is an interesting *dramatis personae*. I will also show you how times have changed and how differently we work in the current era. The speciality has seen truly disruptive change, particularly the introduction of cardiopulmonary bypass (the heart-lung machine), intensive care, computing and imaging as well as dramatic improvements in materials science. These changes encompass many of the reasons I chose this subject for the them of my lectures and provide a prologue to later talks.

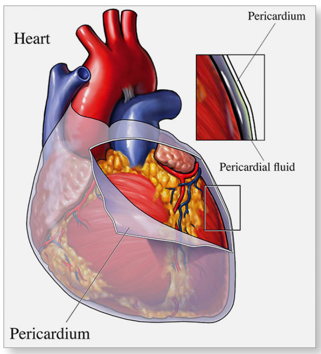
Heart surgeons might have become some of the medical superstars of the late 20th century, but this was not how they were perceived by a19th Century superstar. **Theodor Billroth** (1829-1894) is usually regarded as the father of modern surgery, and these were his views:

"A surgeon who tries to suture a heart wound deserves to lose the esteem of his colleagues"

"Performing an operation to the heart is tantamount to an act of prostitution in surgery or surgical frivolity".

http://www.aerzteblatt.de/archiv/54013/Medizingeschichte-Herznaht-wider-ethische-Bedenken

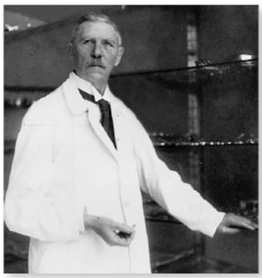
Fortunately, some were not put off by the derogatory Billroth, and, indeed, surgery on the heart started with the repair of stab wounds, following the predictions of John Bingham Roberts in 1881 *"The time may possibly come when wounds of the heart itself will be treated by pericardial incision to allow extraction of clots and perhaps to suture the cardiac muscle."*



The first published reports(6) being in the late19th century from **Henry C Dalton** in St Louis in (1891). Actually, he only stitched a tear in the pericardium, but that had filled with blood and was compressing the heart, so it was just as effective in saving life.

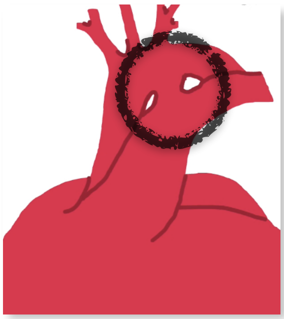
Hale Williams did the same in Chicago in 1893, and in 1906 **Ludwig Rehn** of Frankfurt reported(8) 124 cases which had been repaired in Europe in the 1890s, with 40% survival, remarkable for the time.

Ludwig Rehn (http://www.aerzteblatt.de/bilder/2007/01/img122602.jpg)



Surgery on the heart and its immediate branches was being researched at around that time. **John Munro** demonstrated in 1888 (on an infant cadaver) the feasibility of closing a persistent ductus arteriosus (PDA), but did not want to carry it out on the living because of ‘uncertainty of diagnosis’(9). An urgent ligature closure of an infected PDA was carried out by **Strieder** in Boston in March 1937(10), but the patient died of overwhelming sepsis 4 days letter. However, it was not until 1938 that **Robert Gross** closed a PDA by division and separation(11). Gross usually gets the credit for being first for some reason.

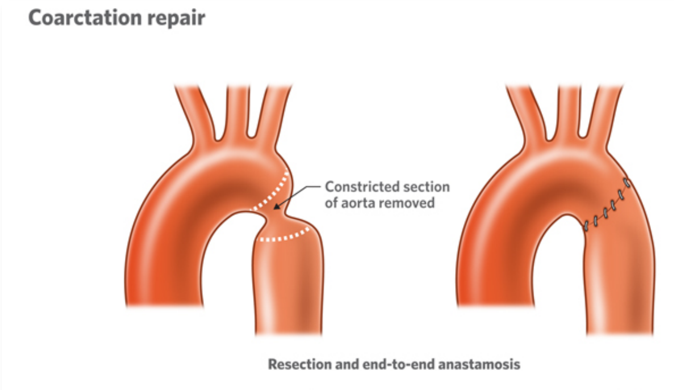
The ductus arteriosus usually closes immediately after birth, but if it persists blood can flow from left to right into the lung arteries causing heart failure, or increase the risk of infection at a site of turbulence. You might remember seeing the PDA form during the animation of the development of the heart by **Jacob and Matt** in my first lecture, highlighted here between the aorta and pulmonary artery:-



Robert E Gross
(http://www.fa.hms.harvard.edu/docs/memorial_minutes/images/gross_robert_e.jpg)

Gross, working in Boston, closed the ductus with a combination of sutures and ligatures, but did not enter the heart. This was ***extra-cardiac surgery***, performed through the left side of the chest. It was still brave though. The aorta and pulmonary artery each carry about 5-8 litres of blood per minute in an adult. He did it when his boss, Ladd was away (real *chutzpah* said Bartlett, later(5)). He was fired on Ladd’s return, but waster appointed as the Ladd professor at Mass General. Amazingly, Gross was blind in one eye, because of a congenital cataract; depth of vision is extremely important to the surgeon.

In 1944, after learning PDA ligation, **Clarence Craaford**, a meticulous perfectionist, in The Karolinska Institute in Stockholm reported the repair of aortic coarctation in an 11 year old boy.

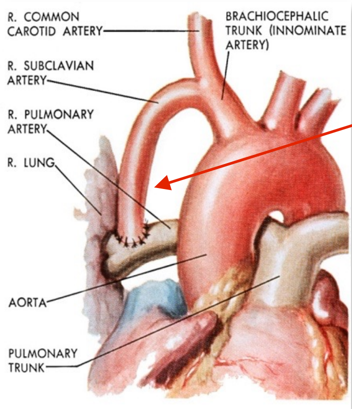


Coarctation of the aorta is a congenital narrowing of the aorta (the main vessel leaving the heart) which can result in severely high blood pressure and heart failure. Craaford clamped the aorta above and below the narrowing, chopped out the narrow segment and stitched the two ends together. Routine nowadays, but in those days patients were sicker, further along in the course of their disease, older than we operate on them today and the sutures and needles that Craaford had to use were primitive. Instruments were large and clumsy and light sources were, by our standards, dark. These were amazing people; but not without 'issues'! Gross , a hugely competitive man, thought that Craaford had 'nicked' the idea for the operation when visiting his lab in Boston, and irascible and jealous as he was, never quite forgave Craaford(5).

The 1940s were, of course, dark days, dominated by the War. And as always in times of war, there are excessive numbers of injuries, and concomitant rapid advances in medical practice, especially in surgical techniques and skill. **Alfred Blalock** (1899 - 1964) was indirectly responsible for saving many lives during the second world war because of his research into traumatic shock whilst at Vanderbilt in Nashville. He found that blood and plasma transfusions could mitigate the effects of traumatic haemorrhage , reducing both death rates and organ damage. He suffered from TB at this time and so, rather than having to fight, was able to carry out more research in the lab, from 1941 at Johns Hopkins in Baltimore, this time studying the effects of high blood pressure in the arteries to the lungs, pulmonary hypertension. His experiments required him to join the high pressure artery to the arm (subclavian artery) directly to the low pressure artery to the lung (pulmonary artery). This was very difficult surgery in its day, and Blalock was helped enormously by his gifted technician, **Vivien Thomas**, the son of a slave, who was not even allowed to enter the front door of

Dr. Alfred Blalock Vivien Thomas

PHOTOS BY BY THE ALAN MASON CHESNEY MEDICAL ARCHIVES OF THE JOHNS HOPKINS MEDICAL INSTITUTIONS

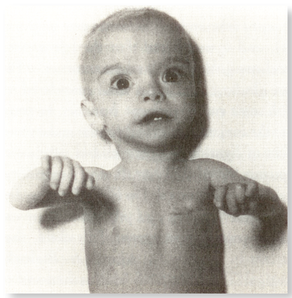


The artery to the right arm, turned down to join the artery to the right lung

the hospital, yet whose talent Blalock recognise and insisted he come to Baltimore with him. Their story was recently made into an Emmy award winning film called "Something the Lord Made", starring Alan Rickman and Mos Def (HBO 2004).

Whilst these operations were initially performed to *create* a disease (pulmonary hypertension). **Helen Taussig** (1898 - 1986), originally form Boston, who moved to Baltimore to work as a paediatric cardiologist, realised that if something could be done to get more blood to the lungs of blue babies then they might survive for longer, and she approached Blalock with her idea, having previously floated it to Robert Gross who wanted nothing to do with it, saying he had enough trouble closing a duct (the communication between aorta and pulmonary artery) and did not want the bother of having to build one . It was, however, an idea of its time, when skill and initiative merged. Helen Taussig, incidentally, was deaf and dyslexic, and a marvellous role model.

The first patient operated on was Eileen Saxon, aged 15 months, shown here on the right (*image courtesy of Professor Luca Vricella, Johns Hopkins University, Baltimore)*.



William Longmire, who assisted Blalock at that operation, described(12) the child thus:-

".. On evening rounds, we arrived at the crib of this fifteen-month old baby… I was immediately astounded by the deep cyanotic appearance of the child, much more cyanotic than any patient I had ever seen before: the lips were a deep, dark blue .. The face was suffused with dilated veins, the conjunctiva almost purple." In a later letter to Naef(4)he described the operation in equally compelling language:- *"At operation we lacked all the modern vascular instruments and really had very little but the professor's determination to carry us through the procedure. With the extremely thin wall of an extremely small pulmonary artery I marveled at Dr Blalock's determination in completing this first anastomosis, certainly the most difficult I have ever seen".*

Switching Arteries Sidetracks Blood and Oxygen to Otherwise Starved Lungs

Saving Our Doomed 'Blue' Babies

The "Blue" Babies' Blood Lacks Vital Oxygen Because the Artery (1) From the Heart to the Lung Is Constricted. By Severing an Artery of the Arm (2), Tying It Off (3) and Attaching It to the Lung Artery (4) the Constriction Is By-Passed.

By Robert D. Potter
Science Editor

A WOMAN physician's courageous research and imagination, and the skill of one of the world's great surgeons have combined to bring hope that many "blue" babies—hitherto considered doomed to early death—may be saved.

These babies are blue because they are suffering from a lack of oxygen in their blood streams, in a condition known as cyanosis. The artery from their heart to their lungs is so constricted that their blood never gets oxygen to make cheeks rosy.

Their lips are blue. Their toes are blue, and they can walk only a few feet without exhaustion. Doctors used to give them only a few tortured years to live.

But now medicine can give hope . . . and more . . . for since Nov. 20, 1944, Dr. Blalock, Professor of Surgery at Johns Hopkins University in Baltimore, has been conquering the "blue" baby malady by rerouting an artery from the arm and making it carry blood to the lungs where it can receive its vital oxygen.

Nearly 70 operations have been performed on "blue" babies. In many cases almost miraculous recovery has come.

It is Dr. Blalock's fingers that wield the knife in the delicate operation that exposes the heart and transplants its vital arteries. But behind the brilliant operation he has perfected are years of painstaking research by Dr. Helen B. Taussig.

Daughter of the late Prof. F. W. Taussig, world-famous Harvard economist, Dr. Taussig had watched "blue" babies come to her heart clinic at Johns Hopkins Hospital.

In many cases she discovered that the artery leading to the lung from the heart was narrowed so that an insufficient supply of blood was reaching the lungs to receive its vital oxygen. Dr. Taussig reasoned that a surgical operation might be able to short-circuit the constriction and sidetrack blood into the lungs. On paper, when the diagram of the art- the blood would pick up its life-giving oxygen. Then it would go back to the heart again to move outward through the body.

But could it be done? It is one thing to have a plumber re-arrange a piping system and something quite different to lay bare the human heart, sever one of its main arteries, splice it to another main artery and sustain life in the patient in the process. Dr. Blalock said he would try.

Since the pioneer attempt the operation has been largely successful, although it is one filled with danger. Among the first 70 patients, 14 died. The odds are 5 to 1 for success.

Now that the news of Dr. Blalock's operation is known through the country the list of patients grows daily. Blue little Bonnie Stewart of Florida, daughter of a daddy killed on Saipan, went to Baltimore with her grandmother. Today Bonnie walks and plays like other children.

The case of six-year-old Mike Schirmer—the boy with the "itchy zipper"—shows what can be done.

Six-Year-Old Mike Schirmer of Baltimore Could Walk Only Five Feet Without Resting Before the Operation. He Shows His "Itchy Zipper"—the Incision for the Operation.

Little Bonnie Stewart of Florida Is Another of the 70 Children Saved by the New Johns Hopkins Surgery.

Mike's "itchy zipper" is the healing incision over his heart where Dr. Blalock went in to do the operation. But let his mother tell his story:

"Michael could only walk five feet and then he'd have to squat down on the sidewalk and rest.

"I had to wheel him everywhere. Strangers would stop his carriage and was no hope that Mike could grow up. But then came new hope, for Dr. Mansderfer told us about the operation of Dr. Blalock.

"They took him to the operating room and brought him back two hours later. It was a miracle.

"After only two weeks of convalescence he came home and he has been on the go ever since. If anyone wants anything he'll run and get it. He's up and down stairs 75 times a day. He climbs on bureaus and tables just for the joy of jumping off. He wears me out. But I love it."

The Blalock-Taussig operation is not a simple one. It takes from an branches of the pulmonary artery (to the lungs) are two large blood vessels. One connects the heart and the arm, the other the heart and the head. Dr. Blalock chooses the most convenient—usually the arm artery—and severs it. One end is clamped off and the other closed permanently.

The end nearest the heart is then spliced to the nearest branch of the pulmonary artery. The clamps are removed and the blood that would ordinarily flow to the arm goes into the lung. There it becomes enriched with vital oxygen and the baby's blue lips quickly begin to turn red.

What happens to the arm?

Nature has provided other blood vessels which take up the blood load

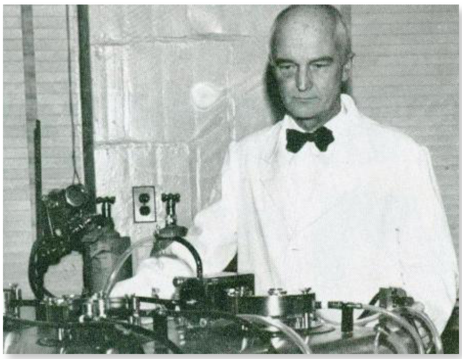
The operation revolutionised the lives of children who otherwise would have died a rapid and miserable death; blue, exhausted and short of breath, and terrible for their families to watch.

The Blalock-Taussig shunt as it became known attracted a great deal of media attention, and spread rapidly around the world. But whilst it was a help, a palliation, it was not a cure. The underlying problem within the heart had effectively been bypassed, and patients remained blue, although less so, but had a much shorter life that normal.

http://www.medicalarchives.jhmi.edu/tausbio.htm

As I explained in my last lecture, the most important heart problems which you could be born with were *inside* the heart. Whilst Blalock, Craaford and Gross were developing techniques which did significantly help children, two other parallel pieces of work were going on which enabled modern open-heart surgery.

In 1931, at the Massachusetts General Hospital, one of the most important pioneers of my field, **John H Gibbon**(13), who came from an 'old money' Philadelphia medical dynasty, watched Edward Churchill remove a blood clot from the main lung artery of a young woman in 'snatch and grab' operation lasting 6 minutes 30 seconds. The lady died, and her, what he thought unnecessary, death drove Dr Gibbon to develop a mechanical method to substitute temporarily for the heart and lungs to allow the inside of the heart to be operated on for longer periods. He (and **his wife Mary**, who was Dr Churchill's technician) began work which, over the next 20 years, led to the development of a heart-lung machine permitting open heart surgery in humans. It is hard to find anyone with a bad word to say about Gibbon, who seems to haven erudite and charming. But there must have been some element of ego about him. Bernie Miller was working in his lab and claims to have modified the machine enormously, and he and the other lab rats remained bitter that their work was never recognised by Gibbon. Perhaps this just reflects the importance of figure head leadership, prevalent at the time.



John Gibbon and an early heart lung machine

This was truly disruptive technology, as **DeBakey** said(14) *"blasted open the door that had been locked for centuries against any medical therapeutic intrusion in to the cardiovascular field".* Prior to the development of heart surgery, and into the 1950's, the physician caring for children with congenital heart defects had little to do but make death as comfortable as possible. Many described a feeling of helplessness.

After demobilisation in 1945, a young Canadian called **Wilfred Bigelow** spent a year at Johns Hopkins, watching the pioneers of closed heart surgery, he wrote later(15)

Wilfred Bigelow (1913-2005)

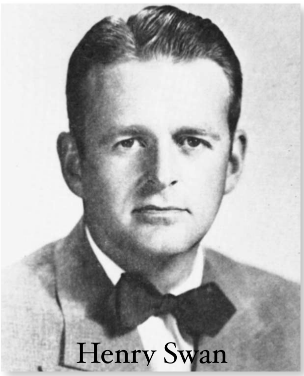
"While watching operations performed with the heart beating forcefully, I realized that surgeons would never be able to cure most heart conditions unless they could atop the circulation of blood through the heart, open it, and operate in a bloodless field under direct vision. At that time the heart-lung pump was not yet practical. Then the inspiration came: "One night I awoke with a simple solution to the problem, and one that did not require pumps and tubes -- ***cool the whole body, reduce the oxygen requirements, interrupt the circulation, and open the heart.***"

Bigelow and his group did his research into hypothermia and hibernation on groundhogs (which hibernate) to start with, and was able to cool them to 5°C and operate on the open heart for two hours. Non-hibernating mammals did not tolerate such low temperatures, or the rewarming which followed but, by 1950, he was able to operate for short periods on dogs at warmer temperatures with a 30% survival.

Techniques (beyond the scope of this lecture) were refined and in 1952, **John Lewis and Richard Varco** closed an atrial septal defect ASD (the simplest internal ‘hole in the heart’) under hypothermia in 5½ minutes. Bigelow had never heard of Lewis(5), who had never contacted him, and was naturally disappointed. But Bigelow was modest and quiet, with rather Scottish-Presbyterian morals, and not possessed of the competitive ego of so many of his peers.

Illustratiuon of the atrial well methode to close ASD as described by Robert Gross in 1952.

Meanwhile, Gross in Boston was trying something else for this hole. Here is the method he employed, the so-called atrial well technique, operating essentially blind in an open well of blood; you don't have to be a surgeon to think through the risks of this procedure.



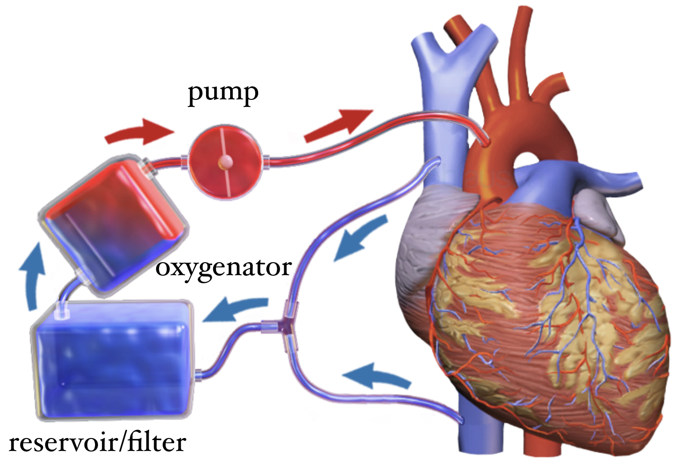
Henry Swan in Colorado preferred hypothermic techniques, starting in 1953 and in a few years had built an extraordinary experience of hundreds of cases with low mortality.

http://www.heartviews.org/articles/2008/9/3/images/HeartViews_2008_9_3_128_63765_sm6.jpg

So now surgeons had the basic tools to allow surgery on the open heart. But these were still very contentious techniques, and although lives were saved ,complications were many. The alternative to surgery was death in most cases, but diagnostic techniques were primitive, decisions were still difficult and the teams involved had to be driven, brave and supported by very courageous patients and their families.

These problems are well illustrated by Gibbon’s early experience. Dr Gibbon did his first human operation in February 1952, using his new heart lung machine machine on a 15-month-old girl with an alleged atrial septal defect. The then current diagnostic techniques were not precise enough, or themselves very risky. Unfortunately, this little girl did not have an atrial septal defect but rather a left-to-right shunt through a large patent ductus arteriosus. Sadly, she died on the operating table. Such an experience would have stopped many there and then, but Gibbon and his team tried again on 6th May 1953, on an 18 year old with a clearer diagnosis of ASD. She had 26 minutes of bypass, her hole in the heart was closed, she did very well and went home within 2 weeks. This operation may have changed the world, but for Gibbon, things did not go so well. His next two patients, both aged 5, also died on the operating table and he never again did open-heart surgery.

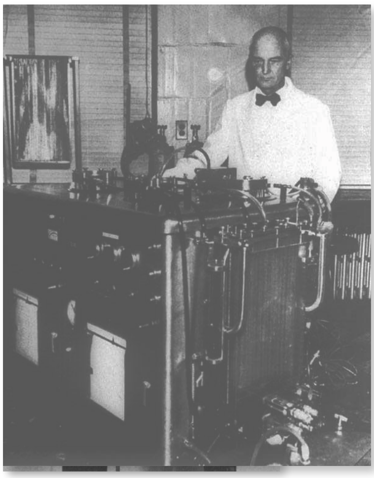
I want to take a moment to describe some of the problems Gibbon and his wife Mary faced.



A heart lung machine needs piping to connect it to the patient, a reservoir to hold blood, a pump to replace the function of the heart, some means of adding oxygen and removing carbon dioxide from the blood and all this must work without the blood clotting and without pumping any air into the circulation of the patient.

Of course, the Gibbons built on the work of others, including Carrell and Lindberg, but they had innumerable problems to solve in creating each component so that both blood and patient would not be damaged. Their attention to detail is legendary, but they had no big grants and there was, as a result, an element of 'Heath Robinson' about both the way they worked and the equipment they built. Relationships with industry, and especially IBM in those early days were vital in ensuring success.

Experiments were difficult, long-lasting and energy sapping, as Gibbon managed to combine clinical work with his research. And they also had to find enough animals on which to develop there techniques. How times have changed! Jack and Mary saved money by walking about the Boston streets at night securing cats without expense(16). As Jack put it, "*I can recall prowling around Beacon Hill at night with some tuna fish as bait and a gunny sack to catch any of*

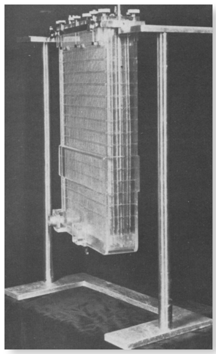


those stray cats which swarmed over Boston in those days. To indicate the number the S.P.C.A. was killing 30,000 a year."

The first heart-lung machines were massive, with roller pumps designed by **Michael deBakey**, and with huge oxygenators comprised initially of rotating drums to 'film' the blood in oxygen, and later is a series of sheets.

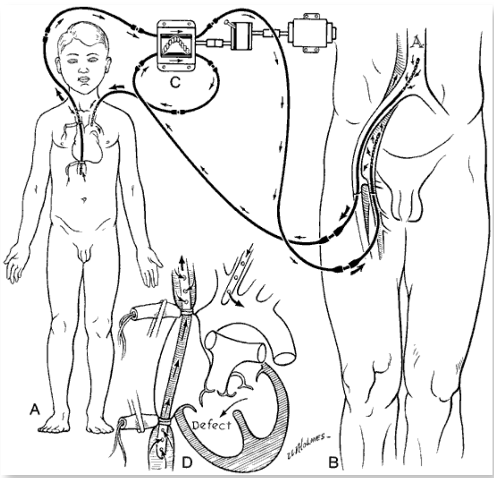
Gibbon's first rotating drum

Gibbon's later screen oxygenator

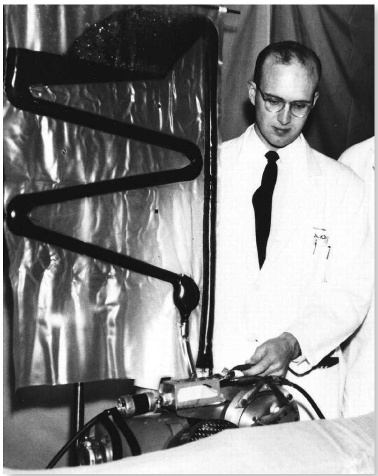


The final heart-lung machine was massive, and needed several technicians to control it during surgery. For Gibbon to have operated whilst still worrying about the safety and effectiveness of his machine, must have added significantly to the stress of his initial procedures. And the fact that he gave up is not wholly surprising. However, the moratorium on open-heart surgery using the heart lung machine that he initiated, did open the door to another remarkable member of our cast of pioneers.

C. Walt Lillehei (1918-1999). Lillehei, active in Minnesota, was also driven to make open heart surgery work, and would not be held back by such attitudes. Afterwards often described as '*maverick*', with bizarre sartorial and colour sense '*like a bookie at a race track*', Lillehei decided to carry out open heart surgery on a Gregory Gliden, using his father as an oxygenator and a pump. This was called *cross-circulation.*



Cross-circulation was described by a contemporary, but anonymous critic, as '*one of the few operations which could have a 200% mortality risk*'. However, he actually did 45 of these operations with a 62% survival at a time when the published survival using a heart-lung machine was 6%. He had conquered the world and as Melrose See below) later put it "*he wanted you to know it*"! Although Lillehei had huge resilience and had come to be able to deal emotional with a 'bad run, he soon realised that cross-circulation was not the way to go. He and others at Minnesota, particularly **John Kirklin (1917-2004)** continued to research the heart-lung machine and modified the apparatus considerably to make it safer and better understood. Both he and Kirklin worked closely with paediatric pathologists to dissect the hearts of poor babies who died, better to understand what they had to deal with.

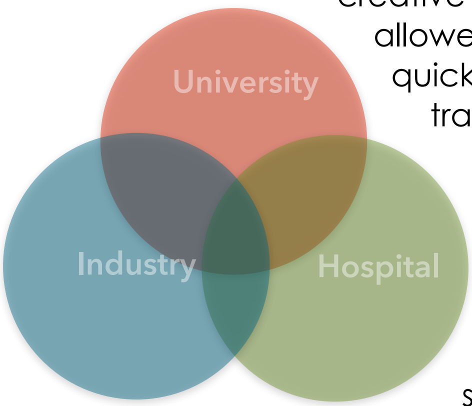


It required two further key advances to precipitate the avalanche of discovery and innovation which followed. These were, firstly, the bubble oxygenator developed by **Richard DeWall** and Lillehei which could be mass-produced, making open-heart surgery possible world-wide. Lillehei had close relationships with industry and with the University, again stimulating an early market in these devices.

The second advance was the discovery that the heart could be arrested by infusion into the coronary arteries of solutions containing potassium(17), and restarted pretty well at will by reperfusion with ordinary blood. This was called **cardioplegia**. This discovery was made by a South African, **Denis Melrose (1921-2007)** at the Hammersmith Hospital in London, working with several British surgical innovators including **Bill Cleland** and **Hugh Bentall**. The solution they created was based on potassium citrate, and whilst that was superseded in later years by much safer and more effective solutions, it is their pioneering work that, with hypothermia and the heart lung machine gave surgeons the time they needed to create and carry out complex repairs in side the heart.

http://wpps-centenary.org.za/wp-content/uploads/2013/10/Denis-Melrose.jpg

We have heard a great deal about these early workers in the field. They share many characteristics. They were **driven** to make heart surgery possible both for reasons of simple humanity (as evidenced by Gibbon, above) and by deep curiosity and a love of solving problems. Drive, intelligence and focus were not enough though. They worked, for the most part, in institutions also committed to innovation and development, and with good and creative relationships with manufacturing industry which allowed their ideas togged to market remarkably quickly. Hospitals in the USA have long had a tradition of being research hospitals, rather than hospitals which do some research. The entrepreneurial spirit, the diligence of individuals and fully integrated academic and health systems combined to create maximum effect and successfully develop the heart lung machine, taken to market with speed. That was also the prevailing spirit at the Hammersmith at the time.



In the UK in 2014, there is often conflict between the clinical (and productivity)demands of the NHS and the academic requirements of Universities, each having separate performance metrics, often competing directly and destructively for the clinical academic's time. These differences have become exaggerated as a result of the current austerity package and reduced or, at best, static funding of public services.

The curious mind is always present, but carving out time, space and resources to develop ideas in the modern NHS can be very difficult. The close relationship between the identification of a clinical need and the ability to work directly to solve that problem in an almost directly adjacent lab is built in to much American training, but can be difficult to achieve in the UK today; research is often 'nodded to' but the time it takes is not respected, and, in our target-obsessed culture, is often perceived as actually in competition with the needs of patients. If those attitudes had prevailed in the 1940s and 50s in the institutions in which our pioneers worked, one wonders whether open heart surgery would have been developed so effectively or so early.

The personalities of the pioneers in my field are often commented upon in the various histories and published biographies. Often coming from scientific backgrounds rather than just medicine, they were honed in laboratory technique and research methodology. They were obsessed by detail and goal-orientated. They were also brave; and the word 'maverick' comes up more than once, especially in relation to Lillehei, and cross-circulation. They were certainly charismatic, easily motivating others by their energy and skill, but some achieved greatness by calm and control in the operating room (Kirklin, Blalock) and others created more theatre around them, indeed Lillehei's operating room was actually described by one observer as a circus.

We must remember that the alternative to what they were doing was, usually death, and despite the terribly high attrition rate at the beginning, they kept going, always aware of the 'long game'. Some were frankly sensitive, notably Gibbon, others clearly thick skinned. All had to show significant resilience to survive both the brickbats of their peers and the emotional trauma of losing patients that they had come to know well. They were using equipment they designed, and often manufactured, and techniques they developed. They must have felt guilt and suffered personally but, largely, kept going. They were supported by the environment in which they worked and especially the holy trinity of hospital, university and industry working in close harmony. Individual patients are the best at motivating research staff; humans love to have a problem to solve, and a good reason to solve it.

With the development of the heart-lung machine, the leash had been let off the cardiac surgical community, and the next four decades saw the un-precedented development of accurate diagnosis, surgical technique and new operations, as the physiology of complex circulations came to be modified. There was parallel development in everything associated with the discipline. Anaesthesia improved dramatically. Equipment became better, transistors replaced valves, chips replaced transistors. Plastics replaced rubber; silastic appeared, and surfaces became smoother. Monitoring of pressures e.t.c. moved from column of fluid to direct transducers, and monitors themselves changed from smoked kymographs through oscilloscopes to flat LED screen devices with built in memory. Tubing and pumps became smaller, and bubble oxygenators were replaced with safer, smaller membrane oxygenators.

Syringe pumps replaced drip sets, and so drugs could be given by much more accurate, weight-related infusions. Operating lights became brighter, and surgeons could wear light-weight magnifying glasses with good optics, and headlights to permit them to see in the deepest or smallest cavity. And finally instruments just got better and better, finer and finer, more and more precise. All conspiring to make it possible to operate on smaller and smaller babies, closer and closer to birth.

With the development of ultrasound which I described in my last talk we were able to introduce quality control in the operating room and permit longer term accurate follow up of structure and function of the heart, giving feedback to aid our decision making.

There was obviously a great deal of innovation, and a significant amount of teamwork was necessary. The relationships between the cardiologist (making the diagnosis), the anaesthetist (caring for the child during surgery), the perfusionist (manning the heart-lung machine), the cardiac morphologist (teaching all the staff about the morphology of the heart) and the amazing nurses who cared for these patients were crucial to success. The best units had the best teams. But, until the 1990's, the units, and often the developments, were almost always surgeon-led.

Why should this have been the case? *Firstly*, none of these conditions could have been repaired without a surgeon, and all the initial developments required someone to be 'brave enough to try'. *Secondly*, surgeons had (and have) to have enormous confidence in their ability, both technical and intellectual. *Thirdly*, the performance a unit, in the eyes of others, was equated to the performance of the surgeon. Few others were ever mentioned in the press. And heart surgeons were not known for hiding their light under a bushel. They were often on the front pages, were seen as highly charismatic and, to people like me, were a big magnet to pull me into the profession.

Just for a moment imagine what it is like to operate on a small baby. A baby that someone loves. As the surgeon you have to;

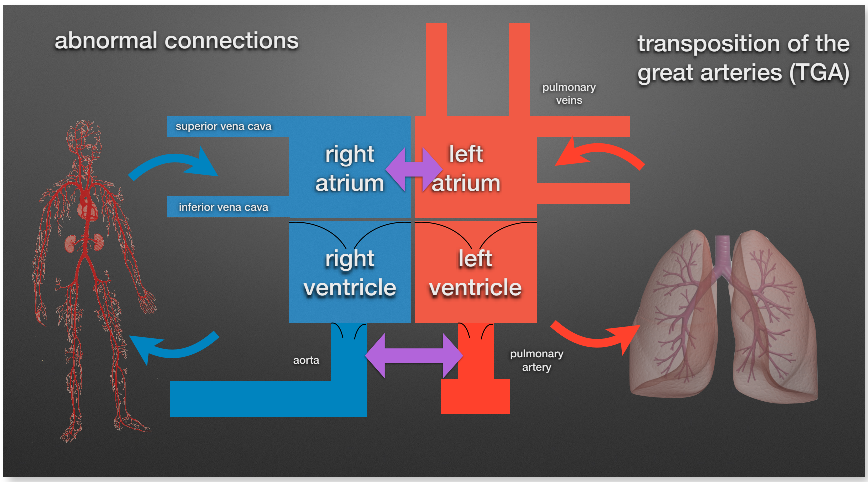
- know and understand what is wrong with baby, *and* in 3 dimensions (the imaging is often in 2 dimensions, and in different projections; the surgeon has to be able to 'reconstruct' these views in her head to visualise the true 3-D appearance, seen from their own perspective).
- be aware of what they want to do surgically, and have back up plans if it proves impossible or something bad happens.
- be able to explain it to those around them
- orchestrate the process in the operating room; leadership was assumed
- AND have the confidence to make that initial incision in the skin. Anywhere else, such a cut would be assault. You have to be sure that you can put it all back together again, and in the context of congenital heart disease, understand the consequences for the child, its circulation and its family of the revisions to the circulation you are about to make.
- deal with all of this at the same time as having the technical skill to work inside a heart the size of a walnut, composed of the most delicate and fragile tissues, damage to which can seriously harm or kill the child.
- Finally, you have to operate 'against the clock'. there has always been a limit to the tie the heart can be protected by

How much of this is science, and how much art? There is clearly a strong foundation of science as we have seen, but there is also an element of craft and creativity, and dealing with the surprises thrown up by poor or incomplete diagnosis truly requires, and tests, both. The best surgeons I have seen and worked with were also the best cardiologists, the deepest thinkers and the best technicians. Technical skill is how surgeons describe the physical elements of their craft to each other, but watching a great surgeon reveals more than that; it is beautiful to watch. Great surgeons are efficient in their movements, gentle with tissues, unfazed by the unexpected and in control. There is a balletic grace about their surgery, that combination of skill, training and movement that makes art out of craft.

It is not surprising in retrospect that the leadership needed in the operating room frequently translated into surgical leadership of the wider service in which these surgeons worked. To the wider world, and certainly to the management of the hospitals and the media, the surgeon's name was often equated with the unit, and the reputation of the surgeon was the reputation of the unit. They were interchangeable and even today, especially in the USA, if a unit is not doing well it is often the surgeon that is 'swapped out'.

But surgeons are surgeons, and thus largely α-male (there were few women to start with) and very competitive. As I indicated, new operations were coming thick and fast during the decades after the heart lung machine. They were often eponymous procedures, named (often in genuine peer recognition) after the surgeon who described them. Several of these operations became the yardstick by which other centres and surgeons would judge themselves and others by their success (or otherwise) in these procedure. And a surgeon would not have much 'street cred' if they could not do them. Each generation of surgeons had (and indeed has) its own test-case operation. I cannot do justice to all the surgeons who have made massive advances in the last half century, but there are some that I want specifically to draw your attention, because of their clinical, ethical and, in some cases, political importance.

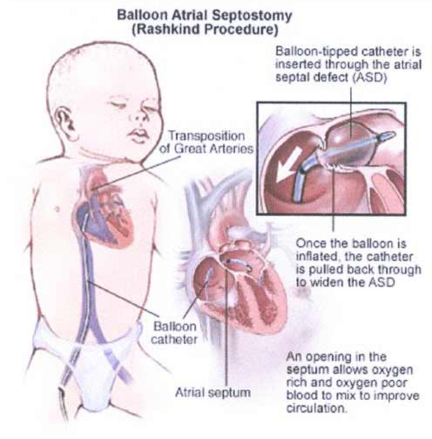
Let's start with surgery for transposition of the great arteries (TGA), the



condition in which the child is born with the two major vessels leaving the heart being connected to the wrong ventricles.

In my first lecture, I described this condition using the above diagram, and pointed out that you could only survive after birth if a way of mixing red and blue blood could be preserved or created after birth. To jump a few years ahead, we can see that ways evolved to create or sustain these sites of mixing.

Making a hole between left and right atria (see a in diagram above) could be done surgically, but a revolution occurred in 1966 when a cardiologist called **William Rashkind**, working in Philadelphia developed a technique which allowed a hole to be created or enlarged by a balloon passed up from a vein in the leg:-

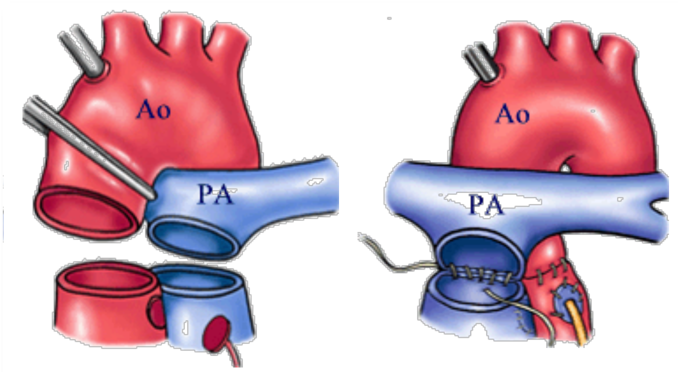


http://users.skynet.be/bbnc/hartafw/Rashkind.jpg

Not only did this save lives, but with it Bill Rashkind created a whole new discipline; *interventional cardiology*. And it had the added advantage of keeping the surgeon in bed for a few hours longer.

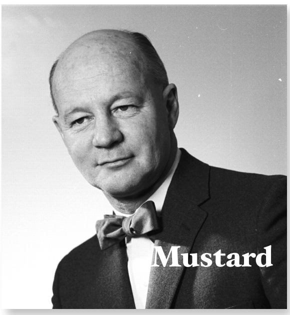
In the 1970's another important development to help me stay in bed came in the form of a drug called prostaglandin, which when given IV could keep open the ductus arteriosus between the aorta and pulmonary artery, allowing mixing of red and blue blood at arterial level (labelled 'b' in the diagram above).

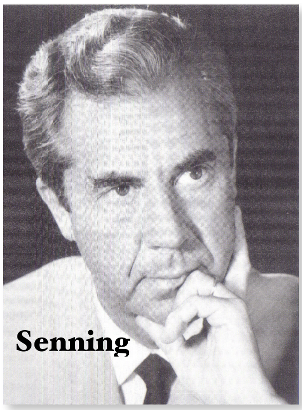
The most obvious way to correct TGA, is to put the vessels back where they should be; theatre leaving the left ventricle, and the pulmonary artery leaving the right. Unfortunately, as in much of surgery for congenital heart defects it is not as simple as that. This is because the arteries delivering blood to the heart itself, the coronary arteries, start immediately above the aortic valve, as shown here;



The coronary arteries have to be moved, from the base of one artery to the other, and at the age the operation is done, they are only about 1mm across. Yet of course, the logic of the operation is clear, and thus it was the first way in which the surgical pioneers tried to fix this otherwise fatal condition.

In 1952, **William Mustard (1914-1987)** in Toronto , attempted to perform such a repair which we now know as **the arterial switch**, switching the arterial outlets of the heart(18). He used a monkey lung as an oxygenator, and only moved one of the coronary arteries, thinking this would be enough to live. He did 6 more that year and none lived for more than a few hours. Another surgeon called Bailey tried it again later that same year, and in 1955 **Åke Senning (1915-2000)** in Sweden had another go. All failed. We can only imagine what that must have felt like for families and surgeons alike, and how much commitment it took for people to carry on trying. Senning was tall and quiet; very self-contained, but a great organiser and planner.





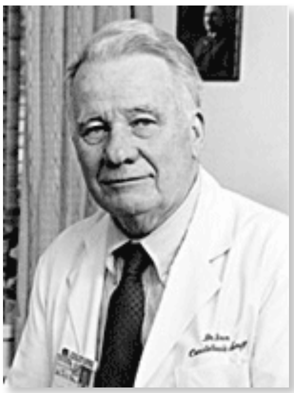
Having failed at switching the **outlet** of the heart in transposition, Mustard and Senning did not give up. Rather they thought rather laterally and designed operations to switch the inlet of the heart, based on the principles outlined in 1954 by Dr Harold M Albert. Senning, in1957 did the first successful inflow switch, in what became known as the **Senning operation**, redirecting the flow of blood inside the atria ingeniously using the child's own tissue. In 1963, Mustard operated on an 18 month old little girl using a patch of pericardium to create a tube diversion in the heart. This was the **Mustard Operation,** which was a little simpler and certainly easier to perform in small children. Indeed, it became the almost universal procedure through ought the next decade until it was realised that the Senning operation, because it used the child's own tissues in the heart, grew with the child and had less late tunnel obstruction. When I started doing cardiac surgery in the late 1970's, the Senning was the procedure that surgeons judged themselves by. It was hard to grasp as a concept and technically difficult to do well. By the mid-1980's, several teams were achieving very low mortalities with this operation (<5%), but two concerns remained. Firstly, there was quite a significant death rate waiting until the child was big enough for surgery (usually 3-6 months of age) and secondly, the child would have to have its right ventricle (RV) pumping blood around the body, into a high resistance, for the rest of its life. And it was already becoming apparent that there was a late attrition rate as the RV began to fail. It was simply not designed to pump blood anywhere except the lungs.

So, despite these very good early results from the Senning operation, several surgeons tried once again to perform the arterial (outflow)switch. **Adib Jatene b 1929** did the first successful arterial switch (the switch) in Sao Paolo, Brazil in 1982. He became Health Minister in Brazil and was recently a presidential candidate. The arterial switch grew in popularity, driven by **Yacoub, Planché, Casteneda, Mee, de Leval** and others. All of whom had themselves made great contributions in bringing down the age of repair towards the neonatal period. However, as the operation spread, early mortality was extremely high, and precipitated important ethical and moral debate. Remember the mortality for the Senning was extremely low. The technical difficulty, size of the patients and this early mortality meant that the arterial switch was the next operation by which units and surgeons earned their spurs. If you couldn't do a switch you were not a 'man'. If your unit was not good at the switch, it was 'second rate'.

Jatene

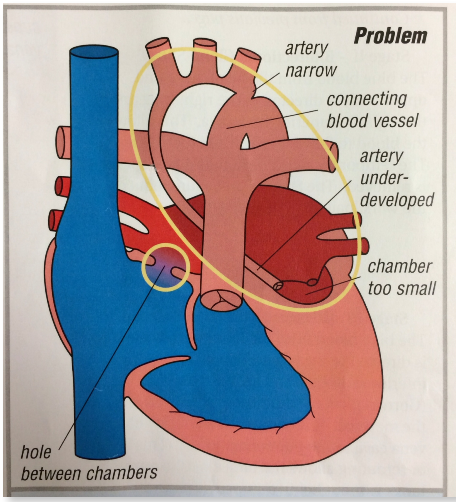
Again, put yourself into the shoes of these early switch surgeons, *believing* but unable to know that the long term results would be better, and having to (a) take the surgical risk of the procedure against their reputation, and (b) 'sell' this high mortality to the families concerned. Their perseverance, and the belief in the principles underlying it has resulted in results for the arterial switch today with mortalities approaching zero in the best units(19).

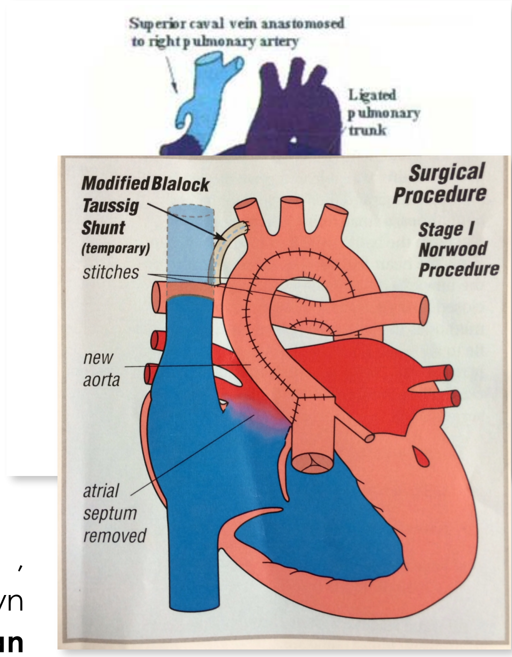
Last time, I demonstrated that certain components of the heart could be missing or severely underdeveloped at birth. Working out how to bypass absent or very small ventricles was on the hit list of most early cardiac surgeons. Dealing with the small left heart was thought to be pretty well impossible until the mid 1980's, but bypassing the hypoplastic right heart was attempted very early. Following animal experiments performed by peers throughout the world, in 1954 **William Glenn (1914-2003)** working at Yale connected the vein (superior vena cava) draining blue blood from there part of the body directly to the artery to the lung, missing out the right heart. This has become known as the **Glenn shunt**, and has formed the basis of modern multi-staged reconstructions of absent right and left heart components.



http://www.averybiomedical.com/images/williamGlenn.gif

Others built on this work in labs around Europe, but in 1968, **Francis Fontan** working in Bordeaux, operated on a young woman whose tricuspid valve was absent, fully diverted all the blue blood returning to the heart around the right ventricle to the lungs. This, of course, became known as the **Fontan operation**. He was trusted to do this by his boss, Broustet, despite it never having been done before(20). The woman did well, and is alive today. The details of that operation were not published, despite its success, and it was not until 1970 that he performed the second operation, subtly different from the first, but importantly the precursor of several variants permitting diversion of blue blood directly to the lungs. There are thousands of people walking around today who were considered inoperable before Fontan was brave enough to try his operation on that equally brave young woman.





Francis Fontan is the epitome of Gallic success. He is also an excellent winemaker, and his sauterne (Chateau l'Ermitage) is legendary in the cardiac world!

His father was Victor Fontan, who led the 1929 Tour de France, but had to drop out after his bicycle failed and he spent a night knocking on doors trying to borrow another.

The last eponymous operation I want to introduce to you today is the Norwood Operation. **William (Bill) Norwood (1941-)** epitomises the pioneering era of the cardiac surgeon. He qualified initially as an engineer, and his understanding of the physics of the circulation, and his utter focus and

commitment led him to work out a treatment for one of the most common, but until he came along, universally fatal congenital heart conditions, hypoplastic left heart syndrome. Bill is also a technical master, he has huge hands (size 8½) but operates with spectacular efficacy and speed, stitching simultaneously with both hands. But as as surgeon he was irascible, intolerant of people with less skill than himself, and idiosyncratic in his care of patients, confident in his own understanding of the circulation. He was greatly helped in the early stages development of the operation by the wonderful Aldo Casteneda in Boston, whose somewhat opposite and very smooth personality helped Bill through the tangle of opposition which followed him around.

The operation was controversial form the start. The Norwood operation refers to the 1st of a 3 stage process (Stage 2 being a Glenn shunt and Stage 3 a Fontan-like procedure). Without it the child dies quickly, but with it, there was no knowledge at the time of whether there would be anything other than prolonged suffering for the children with a very uncertain quality of life for them and their families. Initial mortality was incredibly high, and throughout the world the operation was created with a mixture of admiration and scepticism. Many people, including myself, travelled to Philadelphia, where Norwood had moved to become chief, and learn this operation which could promise life to those who would otherwise be denied it. I can say it was a remarkable experience, in many ways!

There are so many things that can go wrong with the surgery, and it is very technically demanding. It is another operation by which centres and surgeons judged (and judge) themselves. It remains an operation performed in only a limited number of places. But more than any other of the procedures I have listed, the children are very difficult to manage afterwards, primarily because it is necessary to balance the resistance to the flow of blood in lungs and body by the use of drugs, ventilation and surgical skill. Norwood basically did this on his own, utterly confident in his own knowledge, and tolerating little criticism. Indeed, he later moved to Delaware and was finally dismissed from there after multiple law suits and criticism of his autocratic and idiosyncratic style. But elsewhere, people gradually worked out how to manipulate the workings of these children after the surgery, and from initial mortalities >90%, the best units now achieve rates of >5%. It is an operation that exemplifies the move from a the single dominant surgeon to the success of teamwork . As James Tweddell, an excellent surgeon from Milwaukee, put it *"Now, it's a matter of refining the technique … rather than enormous leaps forward. The mavericks like Norwood have gone by the wayside. The field is full of fastidious surgeons who have had to become expert at managing risk"*

These pioneers were remarkable people. They were driven to preserve life, in some cases at all costs. They worked ridiculously hard, without a single nod to working time directives. They expected similar dedication from those around them, and largely got it. But none of them could have succeeded without the supporting environment of their institutions which fostered innovation and development.

I have been very selective in my choice of procedures and surgeons for this talk. The choice is only partly made on importance but also relevance to the ethical issues I discuss next time. It would be wrong not to mention my own mentors in cardiac surgery;- Mike Holden from Newcastle who attracted me to it in the first place; Marc DeLeval from Great Ormond Street who gave me the freedom to operate and showed so many people the importance of self criticism and Aldo Casteneda in Boston, who showed the importance of calm, leadership, a commitment to teaching and embraced the long term goal of correcting heart defects as as soon as possible after birth.

In the end, all of us who operate on the heart are doing it for the child with the problem, however attractive and rewarding is the surgery itself. When I arrived at Great Ormond Street in 1984m there was a photograph of a little girl on the wall of the Portakabin that formed the unit in those days. Underneath it was a caption that read " I was born with half a heart, not half a life". Our job is to rebuild the heart to make that life even more fulfilling.

*Thank you to all my colleagues around the world who have helped me with this, but especially to **Michiel Vriesendrop**, a wonderful medical student from Leiden in the Netherlands who has helped me source some of the material used in this lecture, and been a great source of criticism.*

Martin Elliott

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