

COMMENTS

The Development of Paediatric Cardiac Services in Scotland

Dr Trevor Richens

Department of Cardiology

Royal Hospital for Sick Children

Yorkhill Division

Glasgow

Introduction

Developmental abnormalities of the cardiovascular system constitute the commonest form of congenital abnormality. As such, approximately 8/100 infants are affected by congenital heart disease, the vast majority of whom will now undergo complete repair or radical long term palliation. Since the first open heart procedure for congenital heart disease was performed in Scotland in 1959, paediatric cardiac surgery has evolved from a widespread, geographically separated service into the single, high turnover national centre that exists today. In this comment I will attempt to outline the development of paediatric cardiac services in Scotland and further afield, and highlight some of the major advances that have taken place since 1959.

The Past

Until 1938 congenital heart disease was viewed as an interesting group of problems, whose pathophysiology was being increasingly understood, but no treatment was possible. That year at Boston Children's Hospital a 7 year old child had her arterial duct successfully ligated by Robert Gross¹, for the first time offering a potential cure for this group of patients. Helen Taussig then reasoned that if a duct could be closed it might also be created as an alternative source of pulmonary blood flow. Although initially frustrated she formed an alliance with Alfred Blalock, who performed the first classical Blalock-Taussig shunt in 1944, anastomosing the left subclavian artery to the pulmonary artery of a child with Tetralogy of Fallot.² This was followed in Sweden in 1945 by the first repair of aortic coarctation.

The use of "closed" cardiac procedures spread rapidly. However it wasn't until 1955 that results of the first "open" procedures were published. These initial cases utilised a live relative as a bypass circuit and in Minneapolis this human cross-circulation technique was used to repair 32 patients with ventricular septal defect, atrioventricular septal defect and Tetralogy of Fallot.³ That year John Gibbon in Philadelphia was credited with designing the first extracorporeal bypass circuit, which was modified and used by John Kirklin to successfully treat eight patients.⁴

Surgery on adults and children with congenital heart disease evolved from that point, changing the outlook for an expanding number of patients. Open heart procedures on infants however, remained problematic because of high mortality both before surgery and in the perioperative period. The widespread use of

deep hypothermia and circulatory arrest in the early seventies overcame the problems of using adult cardiopulmonary bypass systems in infants and improved the outcome of open repair of cardiac defects in infancy.

Unfortunately pre-operative survival remained a major problem because of unpredictable arterial duct closure. A number of congenital heart disorders are known as "duct dependent", that is to say survival of the child is dependent on patency of the arterial duct. In some cases this is because the systemic circulation is dependent on duct flow. These include coarctation of the aorta, hypoplastic left heart syndrome and critical aortic stenosis. In other disorders such as pulmonary atresia, critical pulmonary stenosis or severe Tetralogy of Fallot it is the pulmonary circulation that is dependent on duct flow, whereas in transposition of the great arteries it is the mixing between the two circulations that is duct dependent. The cardiovascular stability of children with such condition will collapse upon duct closure, resulting in an extremely sick, often acidotic and hypoxic infant. Prostaglandin E1 was first used in humans to maintain ductal patency in 1975, for the first time providing a way to keep such infants alive and well enough to be considered viable surgical candidates.

In 1959, four years after the first open cardiac procedure was performed in Minneapolis, the first open congenital heart procedure was performed in Scotland. An adult patient with partial atrioventricular septal defect (primum atrial septal defect) was repaired at the Glasgow Royal Infirmary. That patient survived the procedure and in fact went on to have a mitral valve replacement 25 years later.

From that point many different centres in Scotland started to perform both open and closed procedures, a practice that evolved from Scotland's uniquely dispersed population. Covering an area two thirds that of England and Wales with only a tenth of the population, at a time when the transport infrastructure was far less efficient than today, the accessibility of healthcare was a major priority for the service. As a consequence even highly specialised procedures such as congenital heart surgery were performed at local hospitals throughout Scotland.

Over time it became clear that high throughput centres achieved better results and so, following a review of cardiac surgery in Scotland in 1976, the Kaye report was published recommending the centralisation of open cardiac surgery to three centres;

Glasgow Royal Infirmary, Western Infirmary, Glasgow and Edinburgh Royal Infirmary. Despite this, closed procedures such as Blalock-Taussig shunts, coarctation repair and arterial duct ligation continued at many other regional hospitals. Towards the end of the seventies children's cardiac surgical services came under scrutiny resulting in localisation of two centres; Royal Hospital for Sick Children, Glasgow and Royal Hospital for Sick Children, Edinburgh, essentially forming a centralised paediatric cardiac service for Scotland.

Despite these changes in the logistics of the service, development and improvement of surgical techniques continued. Although limited by funding restraints at the time when the need for improvements in basic healthcare provision were high, children's heart surgery in Scotland progressed from straightforward ventricular and atrial septal defect closure to repair of Tetralogy of Fallot and physiological repair of children with transposition of the great arteries. This physiological repair, also known as the Senning or Mustard repairs, was first performed in the early sixties, although it wasn't until the description of palliation by atrial septostomy in 1966 (a procedure that allowed adequate systemic oxygenation pre-operatively) that infants started to get to surgery in a reasonable condition. The widespread introduction of prostaglandin E1 to maintain ductal patency in the mid seventies further increased the number of infants suitable for repair.

Through the eighties the cardiac surgical service continued to develop rapidly. Surgical closure of the arterial duct in premature infants was first undertaken in Scotland in 1980 and the first Fontan procedure in 1982.

Children whose cardiac anatomy dictated they would never achieve a normal biventricular circulation had undergone palliative procedures such as a Blalock-Taussig shunt or pulmonary artery band since the early fifties. Unfortunately, despite these palliations, the workload placed on the single ventricle was higher than normal, and the children remained cyanosed. In 1954 Glenn⁵ had introduced the cavopulmonary shunt, which directed superior caval return directly to the pulmonary artery, bypassing the heart and effectively normalising the ventricular volume load, but leaving the children blue. In 1971 Fontan described the first complete right ventricular bypass⁶ in children with tricuspid atresia. Since then it has been applied to most cardiac abnormalities with only one functioning ventricle, and as such, has drastically improved the outcome for a sizeable portion of children born with complex congenital heart disease. The Fontan procedure closed the atrial septal defect and established continuity between the right atrial appendage and the pulmonary artery, effectively diverting the entire systemic venous return to the lungs through the right atrium. Scotland's first Fontan was performed in 1977 and the patient survived until 1994, ultimately dying of a pulmonary embolism.

New techniques were rapidly assimilated into Scotland from that point on; the first arterial switch for transposition of the great arteries was performed in 1984 giving these children an anatomical as well as physiological repair and a better long term

outlook with the left ventricle in the systemic position. By 1989 it had completely replaced the Senning procedure for these children. The first valve-sparing repair of Tetralogy of Fallot was in 1988, reducing the need for future pulmonary valve replacement in this sub-set of patients.

The 1990's saw ongoing refinement in surgical practice. The Fontan procedure in particular has been modified several times, from the original right atrium to pulmonary artery connection to the introduction of an intraatrial baffle directing blood from the inferior vena cava directly to the pulmonary artery along the lateral wall of the right atrium. Subsequently this has been refined further using an extracardiac conduit to bypass the right heart entirely reducing the risk of late atrial arrhythmias and thrombi.

The major advances in paediatric surgery of the last thirty years have been facilitated by equally important though less dramatic advances in paediatric cardiology. The area of medicine concerning children with heart disease has fostered a unique environment in which both physician and surgeon are critically dependent upon each other to ensure the best possible outcome for the child. Primarily the role of the cardiologist has always been that of accurate diagnosis, and appreciation of the optimum time to intervene, however, this has evolved into one with a heavy emphasis on transcatheter intervention.

Although cardiac catheterisation was first performed in 1929 (by Werner Forssmann who introduced a catheter into his cubital fossa before walking to radiology to confirm its placement) its use did not become widespread until the early seventies. Prior to that diagnosis relied heavily on clinical acumen, electrocardiography and radiology. Paediatric diagnostic catheterisation was first introduced in Scotland in the sixties, and rapidly became the principal technique used to obtain an accurate picture of a child's cardiac anatomy and physiology and so guide treatment.

During the mid seventies, however, cardiac ultrasound began to emerge as a non-invasive diagnostic tool. Although the use of medical ultrasound had been pioneered by a Glasgow group in the mid-fifties, it wasn't until the seventies that cardiac ultrasound, or echocardiography, was clinically recognised as a useful imaging modality. It was again in Glasgow that much of the early work on the interpretation of two-dimension cardiac ultrasound in normal children as well as those with congenital heart disease was pioneered and this led on to the development of Doppler ultrasound as a diagnostic modality for the assessment of intracardiac and vascular flow velocities.

Over a twenty year period these techniques largely replaced cardiac catheterisation as the primary diagnostic tool in children with congenital heart disease. They have proved invaluable not only in initial diagnosis, but also ongoing monitoring of these children throughout all aspects of their care. Indications for echo have now extended to include intra-operative epicardial scans to confirm adequate surgical repair prior to chest closure, and peri-procedural monitoring and guidance during cardiac catheter intervention.

The concept of catheter intervention in children with congenital heart disease was first introduced in 1966 when Rashkind performed the first balloon atrial septostomy. In 1967 the first percutaneous closure of an arterial duct was undertaken, after which the field of catheter intervention expanded rapidly. Atrial septal defect closure, ventricular septal defect closure, balloon valvuloplasty of mitral, pulmonary and aortic valves, and angioplasty and stenting of stenosed pulmonary arteries and aorta have all become standard procedures undertaken in paediatric cardiac catheter labs. Whilst many of these techniques were first tried in the seventies before the widespread use of echo, they have evolved over the succeeding 30 years with vast improvements in procedural technique and device design. Transcatheter balloon pulmonary valvuloplasty and arterial duct closure were introduced in Edinburgh in the early eighties and stents were in use in the pulmonary and systemic circulations by the early nineties. In 1996 Scotland became one of the first countries to use the now universal Amplatzer device for transcatheter closure of atrial septal defects, and in 2000 the first ever HELEX septal occluder device was implanted.

As interventional cardiac catheter techniques have developed, cardiac electrophysiology has emerged as a specialty in its own right. In the eighties techniques were developed to allow mapping of the electrical activity of the heart from within. This in turn facilitated the use of radiofrequency energy to ablate abnormal electrical foci and accessory pathways, providing a non-surgical cure for many children previously dependent on lifelong drug therapy. These techniques were first applied to children in Scotland in the mid nineties from which point the service has developed to include a dedicated paediatric pacemaker and automatic implantable defibrillator service.

The Present

In 1999 the Scottish review of acute medical services was published recommending the biggest change in provision of children's cardiac services in twenty years. After a prolonged consultation period, cardiac surgical services together with interventional cardiology were confined to a single centre in Glasgow, maintaining a smaller non-surgical service in Edinburgh. The move took place in 2000, increasing the surgical and interventional catheter workload in Glasgow by 50% and establishing a unified national children's cardiac service.

Since 2000 Glasgow has operated as the main centre for paediatric cardiac services with Edinburgh functioning as a second, smaller centre with intensive care beds, cardiology inpatient beds, outpatient clinics and a non-invasive cardiac investigation unit. Consultant staff from both units meet on a weekly basis to discuss patients requiring surgical or catheter intervention, and Edinburgh cardiologists also travel to Glasgow for catheter procedures and a limited on call commitment. Cardiologists from both centres do outreach clinics throughout Scotland reducing the distances travelled by patients and their families and maintaining a level of expertise locally. These clinics now extend from Orkney in the north to Dumfries and the Borders in the south.

To further facilitate this local care model and in addition cut down on neonatal transfers, a national telemedicine framework has been developed over the past three years. This expanding network allows the two way transmission of live video, still images and echocardiograms, allowing the cardiologist to view the infant, their chest x-ray, ECG and then watch a live echocardiogram, directing the remote operator as to the views needed and how to obtain them. In conjunction the telemedicine service has been utilised by many other specialties including psychiatry, surgery and pathology and has provided a vehicle by which other hospitals can interact in educational meetings at other centres.

Despite a fall in the actual birth rate, the service has continued to grow as the indications for intervention increase. More children are surviving complex procedures in infancy requiring reoperation or catheter intervention and fewer, if any, children are now felt "inoperable". This has produced both an increase in numbers of procedures now undertaken and also an increase in the levels of complexity involved. Needless to say, cardiac services do not operate in a vacuum and improvements in surgical techniques have only been made possible by parallel advances in anaesthesia, cardiopulmonary bypass and cardiac intensive care.

The expansion of the service has brought with it an increase in facilities and staff. The number of paediatric surgical consultants in Scotland has now increased to three and cardiologists to seven. In addition intensivists numbers have increased as have the number of nurses working in theatre, on intensive care and on the cardiology wards.

Two years ago a specialist paediatric cardiac nursing course was started at Caledonian University. This has proved both popular and successful, and has without doubt raised the standard of care provided to children with heart disease. Courtesy of the British Heart Foundation, the number of cardiac liaison nurses in Scotland has doubled to four over the last two years. These specialist nurses provide education and support to children with heart disease and their families and have swiftly become an indispensable part of the cardiac service. This aspect of the service in particular was highlighted in the Kennedy Report as being one of the most valued aspects of the care we provide.

Currently only two groups of children are transferred outside Scotland for surgery. Children requiring cardiac transplantation are in general referred to Newcastle for treatment, returning to Scotland after transplant for shared follow up. Infants with hypoplastic left heart syndrome are currently transferred to either Birmingham or Guy's Hospital in London for surgery. The three surgical stages of repair are performed at the remote centre however all children receive follow up locally in Scotland. Currently we plan to start surgery for hypoplastic left heart syndrome in Scotland in the next two years, further reducing the need for patient transfer to England.

The Future

The Scottish paediatric cardiac service continues to move forward in many other areas. The development of the hypoplastic left heart syndrome service will facilitate a move to more complex corrective surgery in small infants reducing the number of palliative procedures undertaken. Increasingly a hybrid approach to repair is being considered with the surgeon and interventional cardiologist working side by side to correct cardiac defects through an open chest but without cardiopulmonary bypass.

Interventional cardiology continues to make huge strides forward increasing the number of lesions that can be managed without the need for open surgery and bypass. The applications for paediatric electrophysiology are also expanding. The detrimental effect dysynchronous ventricular contraction has on cardiac performance is recognised as a significant problem in children as well as adults. Cardiac resynchronisation with biventricular pacemakers is in its infancy but promises much for the future.

Non-invasive imaging techniques have surpassed all expectations. The latest generation of cardiac ultrasound machines appears capable of producing 3-dimensional moving images of the heart that can rival those produced by MRI, yet be obtained in seconds at the bedside in awake infants. The potential of this technology is huge not just for diagnosis but also for guiding catheter interventional procedures with near real time 3-dimensional information.

The new paediatric intensive care unit has space for further expansion of the service in years to come, and a new, larger catheter laboratory is planned to facilitate the increasing number of joint cardiology/cardiac surgery "hybrid" procedures being performed.

Conclusion

The Scottish children's cardiac service has both directed and followed changes in the care of this select group of patients over the last 50 years. As the unit moves forward we continue to try and keep at the forefront of the specialty, constantly improving the outcome for children with congenital and acquired heart disease. Advances in foetal cardiology and in care of adults with congenital heart disease have not been mentioned, but these areas have also undergone huge changes and are likely to evolve at least as fast as paediatric cardiac services in the future. Foetuses with congenital heart disease go on to be children and then adults with congenital heart disease. The pathological processes and physiology remain the same although the manifestations alter as the child grows. As such an integrated service allowing a seamless transition from foetus to adult, combining diagnosis, intervention and monitoring is now the long-term goal.

References

1. Gross RE, Hubbard, JP. Surgical ligation of a patent ductus arteriosus: report of first successful case. *JAMA*. 1939; 112: 729–731
2. Blalock A, Taussig HB. The surgical treatment of malformations of the heart in which there is pulmonary stenosis or pulmonary atresia. *JAMA*. 1945; 128: 189–192.
3. Lillehei CW, Cohen M, Warden HE, et al. The direct vision intracardiac correction of congenital anomalies by controlled cross circulation: results in 32 patients with ventricular septal defect, tetralogy of Fallot and atrioventricularis communis defects. *Surgery*. 1955; 38: 11–21
4. Kirklin JW, Du Shane JW, Patrick RT, et al. Intracardiac surgery with the aid of a mechanical pump oxygenator system (gibbon type): report of eight cases. *Mayo Clin Proc*. 1955; 30: 201–206.
5. Glenn, WWL. Circulatory bypass of the right side of the heart, IV: Shunt between superior vena cava and distal right pulmonary artery: report of clinical application. *N Engl J Med*. 1958; 259:117–120.
6. Fontan F, Baudet E. Surgical repair of tricuspid atresia. *Thorax*. 1971; 26: 240–248.