Domain of congenital heart disease: past, present and future†

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Received 19 June 2012; received in revised form 9 August 2012; accepted 12 August 2012

Keywords: Cardiac surgery history • Safe and sustainable • Cardiac education • European Association for Cardio-Thoracic Surgery

INTRODUCTION

At the 2011 European Association for Cardio-Thoracic Surgery (EACTS) meeting in Lisbon, the chairman, Juan Comas was able to thank the past chairman (Fig. 1) for developing the congenital domain and organising the annual meetings and education programmes. Since the inception of the congenital domain, there has been increasing involvement of the domain with the Association of European Paediatric Cardiology, and our American colleagues in North America. This has not only given European, but a worldwide, accent to our meetings.

EDUCATION

Education is the raison d’être for the congenital domain. Lucio Parenzam developed the postgraduate Bergamo course. In 2012, these courses were transferred to our new headquarters in Windsor. There will be one specialist congenital week and separate 1 or 2 day courses on particular aspects of congenital heart disease.

We have representatives of the surgical working group of the Association of European Paediatric Cardiology and are helping to organize the World Congress of Paediatric Cardiology and Cardiac Surgery at Cape Town in 2012.

HISTORY

It is easy to forget the problems of patients with congenital heart disease in the early 20th century. Of course, most infants died before they reached maturity. The survivors were usually restricted to those with severe cyanosis and polycythaemia. The patients with the sequelae of rheumatic fever and patients who had endocarditis for which there was no treatment before antibiotics became available and those with congenital heart problems were the stimulus to develop heart surgery. There are too many medical pioneers to mention all, however, Dwight Harkin from Boston is a good example of experience in operating on the beating heart to remove foreign bodies during the Second World War, which led him to lay the foundation of open heart surgery and the speciality of intensive care in Boston [1]. In 1944, Blalock and Taussig [2] in Johns Hopkins Hospital developed their shunt to palliate patients with cyanotic congenital heart disease. In 1939, the surgical interruption of patent ductus arteriosus was performed in Boston followed by the resection of the coarctation of the aorta both in Boston and Sweden. Many innovative surgical corrective procedures followed, with Mustard [3] publishing his results for surgical management of transposition by atrial baffling in 1964.

In parallel with the surgical developments, heart lung machines were developed, in particular by Gibbon [4], over 20 years or more of development. This innovation allowed intracardiac structures to be examined and repaired under direct vision.

CHANGES OVER THE LAST 30 YEARS

There has been a major increase in surgery for acquired cardiac disease, with complex reconstructive procedures and revascularisation methods. More recently, minimally invasive programmes have developed together with transcatheter placement of pulmonary and aortic valves. These procedures will have a major impact on surgical programmes.

In the congenital programmes, the paediatric interventionalist can now repair many of the more straightforward cardiac lesions such as atrial septal defect, persistent ductus arteriosus, ventricular septal defect and can open up valves and stent arteries that would otherwise require major open heart surgery. In our own programme in Birmingham, this has meant a reduction in case load by approximately 50% by removing these procedures (Fig. 2).

INTRODUCTION OF NEW COMPLEX SURGICAL PROCEDURES

The numbers of congenital procedures have been maintained because of the introduction of complex repairs that were not previously possible. The major change has been in the increase in surgery for univentricular heart, predominantly the
hypoplastic left heart syndrome. In the 1980s, Bill Norwood of Boston Children’s Hospital pioneered the three-stage Norwood programme for the palliation of hypoplastic left heart syndrome. [5]. Figure 3 shows the workload at the Birmingham Children’s Hospital (mirrored by data in other centres) created by this univentricular programme. Now, some 80% survive to 1 year of age. However, they are left with the Fontan circulation, which has long-term complications.

Other more specialist complex repairs include primary surgical correction for pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries popularized by, in particular, Hanley’s unit in San Francisco [6].

Congenitally corrected transposition is another challenging condition and double-switch procedures have now become standard for these patients [7].

In the 1980s, the surgical management of transposition was either the Mustard or the Senning procedure. This was transformed by Jatene et al. [8] with the successful arterial switch procedure first published in 1976. The arterial switch procedure has been a success story for the last 20 years, restoring the left ventricle to systemic circulation.

Paediatric intensive care has had a major impact on perioperative management. Largely developed to support postoperative cardiac patients, it has improved beyond recognition, with haemodialysis, extracorporeal life support and complex ventilation being available for all children.

The majority of patients undergoing congenital heart surgery will survive to adult life, and this has had a major impact on the workload for adult cardiology and surgery with increasing numbers needing surgery in adult life.

QUALITY CONTROL

Outcomes following surgery are now carefully monitored in most centres and countries. The mortality differences, however, are usually not of significance. We now need to focus on morbidity and long-term quality of life issues.

PROBLEMS

The organization and structure of congenital heart units are taxing many countries, in particular in Europe. It is a requirement that we provide a high standard and quality of care with low operative mortality and morbidity. It is generally accepted that the more experienced one is in these complex congenital heart operations, the better the results. Safe and Sustainable programme introduced in the UK to help organize and restructure the congenital heart programmes to get the best results for the patients is something that is to some extent contentious but mostly supported by professional colleagues [9]. The aim is to have a programme that is safe, giving good results for the patients but also providing a suitable environment for mentoring and training new surgical and cardiology colleagues.

However, despite the speciality having really started in the early 1950s and the results having improved over the years, there are still areas of knowledge that need improving. For example, the optimum temperature for cardiopulmonary bypass circuits, the understanding of the bypass inflammatory response, the development of artificial blood products and the best way to protect the myocardium.

The development of viable tissue valves would be a great breakthrough and the future for such valves looks promising [10]. Undoubtedly, new models of more efficient valves will be developed.

FUTURE POSSIBILITIES, EMBRYOLOGY AND CHROMOSOMAL ANALYSIS

Further detailed analyses of the chromosomal make-up and embryology of the heart and vascular structures will undoubtedly help with insight into the causation of congenital heart disease.
When chromatin fragment analysis from the mother is available early in foetal life, this may well impact on the families views about termination of such pregnancies.

**STEM CELL THERAPY**

Stem cells are significantly featuring in many areas of medicine at this time and the hope is that they may enable the development of the patients’ owns tissues for the replacement of part of, or even whole, organs. The aim now is not so much to improve on mortality, which is very low, but to make sure that at the end of the operation we leave the patient with the best possible quality of life.

**MECHANICAL PUMPS AND ARTIFICIAL HEARTS**

At one stage, the mechanical heart programme did not seem so attractive, however, I think the development of miniaturized pumps and small power sources really make it quite optimistically possible to support patients with, in particular, univentricular hearts having undergone the Fontan procedure in future.

**CONCLUSION**

The organization and restructuring of programmes will almost certainly spread, allowing safer and more effective surgery with better training and support facilities for the staff performing this surgery. The early antenatal diagnosis of problems in the foetus would allow the choice for families as to whether they wish to continue the pregnancy or might allow the possibility of early foetal interventions.

Stem cell programmes and the development of artificial hearts and mechanical pumps may well impact hugely on congenital heart surgery in the future.

The growth in the adult congenital programmes reflects the success of congenital programmes over the last 30 years, in particular with the development of complex univentricular programmes. However, this does provide a huge workload for cardiology and surgical services.

At our European meetings and education programmes, we therefore have a responsibility not only to describe clinical outcomes but embrace the knowledge of pure science and medicine together with our colleagues of all persuasions.

**REFERENCES**


