Quo vadis paediatric cardiac surgery?

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Some achievements in the treatment of congenital heart defects are discussed. Special comments are made about the persistent ductus arteriosus, atrial septal defect, transposition of the great arteries and the Fontan operation. The differences and similarities between 'corrective' and 'palliative' operations are discussed. The history of the development of supra-regional centres in England and Wales is described and the current situation outlined. The relationship between the number of operations performed and results is emphasised. Current and future training of paediatric cardiac surgeons is discussed and proposals made for the future organisation of care for children with congenital heart defects. The author speculates about how these problems will be solved in view of the decreasing number of children with congenital heart defects. Impact of treatment on the families of patients with congenital heart defects is also considered.

Surgery of congenital heart defects (CHD) is a young specialty. It was born in 1938 when Gross ligated the first ductus arteriosus. In that year, Tudor Edwards was 48 years old. Surgery of congenital heart defects was developing rapidly in the 1950s and 1960s. In the 1970s there was a period of almost meteoric rise from palliations to repairs, from operations for simple lesions to complex repairs in neonates. More recently, heart and heart-lung transplantation has been developed.

I assisted at my first cardiac operation in Prague in 1962. For me, personally, the next 30 years were like a dream come true. During this time many milestones were reached, many surgical triumphs celebrated. But during the same period we have also begun to realise that some of our surgical triumphs have actually represented tragedies for the families involved. We have learned that, when treating a young child, we must not only be satisfied with its survival, but we must critically evaluate medium- and long-term results. We now know that, under certain circumstances, no treatment may be preferable to complex, often brilliant operations which will only have a short-lived effect.

First, I would like to discuss some of the achievements of the past decades in the treatment of both simple and complex defects. It is now easy and safe to ligate the ductus arteriosus (PDA). It can even be closed by the cardiologist during cardiac catheterisation using an umbrella device. However, the operation for PDA in a premature, very small baby is a different matter. We must use special, small instruments and special techniques. Through a small thoracotomy, instead of ligating the duct, we simply occlude it with a Ligaclip®. The hospital mortality rate, even in premature infants, can be as low as 2%.

Another simple lesion is atrial septal defect (ASD). It is possible to close the ASD on cardiopulmonary bypass with a mortality rate of under 1%. However, it is not only very low mortality but also other factors such as short hospital stay, normal health and good cosmetic results that are important. ASD may be approached through the right thoracotomy in order to achieve a good cosmetic result (1). A small, submammary incision is hardly visible and in a girl will be completely covered by a bra. In future, ASD may disappear from the list of our operations. Recently, cardiologists have developed a technique of closure of ASD with a clam-shell device placed during cardiac catheterisation. The current results are not as good as surgical results, but I have no doubt that this will become the future technique of choice for selected patients.

Transposition of the great arteries (TGA) was not corrected successfully until 1958. This defect has a very unfavourable physiology. The pulmonary and systemic circulations are separate. If there is no mixing of blood between the circulations the baby will die soon after birth. The major breakthrough came in 1958 when Senning (2)
suggested a repair using the redirection of blood within the atria. This operation could be performed even in infants, with a mortality rate of under 5%. There were, however, some late complications, and in recent years this type of operation has largely been replaced by the arterial switch operation (3). In patients with 'simple' TGA, the arterial switch has to be performed in the first 2 or 3 weeks of life. In the best units the survival rate is around 95%. TGA is often associated with other cardiac lesions, such as ventricular septal defect (VSD) and left ventricular outflow tract obstruction. We can repair all associated defects in one operation. The Rastelli operation is our operation of choice. The left ventricle is connected through the VSD to the aorta, the proximal pulmonary artery is closed and continuity between the right ventricle and the pulmonary artery is established with a pulmonary or aortic homograft. Patients lead a virtually normal life after the Rastelli operation.

Not all patients with transposition are suitable either for arterial switch or for the Rastelli operation. An example is a patient with severe obstruction below the pulmonary valve and intact ventricular septum. For this combination of lesions, we have devised another operation (4). A valved conduit is placed between the left ventricle and the pulmonary artery in addition to a Senning operation. As the obstruction cannot be resected, it is thus bypassed.

Patients with one ventricle only remained inoperable for a long time. In 1971, Fontan and Baudet (5) described an operation that at first seemed to be against common sense, but it achieved rewarding results. Since the original description, many modifications of the Fontan operation have been suggested. Most of these are based on the assumption that the right ventricle is not necessary to maintain the circulation. Total cavo pulmonary connection is currently our modification of choice. The operation is suitable for many complex defects. In our series of 152 consecutive patients, the operative mortality was 12%.

Among other major steps forward, one should mention the development of interventional cardiology, extracorporeal membrane oxygenation (ECMO) and ventricular assist devices. For children inoperable by conventional techniques, heart and heart-lung transplantation is giving a new hope.

Many people, cardiologists, anaesthetists, perfusionists, intensivists, nurses, physiotherapists, technicans and others, have contributed to the improved survival of children with CHD. In the 1960s cardiac catheterisation could last 3–4 h. The baby would be transferred to the operating room in a critically ill condition, often cyanosed, acidotic, hypotensive and hypothermic. Today, the diagnosis can be established by two-dimensional echo-cardiography in the referring hospital. Appropriate treatment, for example infusion of prostaglandin to keep the ductus arteriosus open in the cyanotic lesions, can be started immediately. The patient then arrives at the specialised unit in a stable condition and the operation can be performed electively.

As a surgeon, I would like to believe that surgical skill is the most important factor in achieving the best results. However, the organisation of the specialty in the country as a whole may be even more important. Before 1980 there were 41 departments of cardiothoracic surgery in England and Wales. All of them were operating on some children. The hospital mortality for closure of ventricular septal defect in the 1st year of life was 10% in four centres which were operating 60% of all VSDs, but the mortality rate was 40% in the remaining 37 departments. Results of other operations showed similar differences. In view of these findings, the DHSS was persuaded to establish nine Supraregional Centres for infant cardiac surgery and provide them with protected funding. In my view, the system worked well. For example, the mortality of VSD closure in infancy for the whole country decreased from 24% in 1980 to 3.3% in 1992.

In 1992 this College convened a Working Party to evaluate the performance of Supraregional Centres and to suggest further possible rationalisation, because at that time 12 centres were engaged in infant cardiac surgery. The data showed that since the establishment of Supraregional Centres, the mortality rate of operations for infants with CHD had decreased considerably. However, there were still major differences among the centres (6). The mortality rate for all open heart procedures performed in the 1st year of life was 15% in the 'best' centre in 1988 and 5% in 1991. In the 'worst' of the Supraregional Centres, it was 43% in 1988 and 30% in 1991. The area between the two curves in Fig. 1 represents possibly avoidable death.

I have looked at the number of infant cardiac operations performed yearly in various centres. Six centres have carried out less than 100 operations per year, with a mortality rate of 18%. In contrast, there were two departments, each performing well over 100 infant operations per year with a mortality rate of only 6% (Fig. 2).

The relationship between the number of operations and the results was also confirmed in a prospective multicentre study of the arterial switch operation for TGA in the United States. Centres that operated less than ten patients during the study period had 55% mortality. Only two centres operated more than 50 patients, and their mortality was 9% (7). Clearly, volume of work is important for achieving good results.

In 1992 the findings of the Working Party on Infant Cardiac Surgery were submitted to the Department of

![Figure 1. Decreasing mortality rates for open heart surgery in infants between 1989 and 1993. The top line shows the results of the 'worst' of the supraregional centres, the lower line the results in the 'best'.](image-url)
Figure 2. Relationship between mortality rate for open heart surgery in infants and number of cases operated.

Health. In summary, the Working Party "re-affirmed that supraregional funding for neonatal and infant cardiac surgery continues to play a vital role in the high standards of practice that is evident in this field in this country. The attempt to establish similar systems is apparent in other countries. The Working Party endorsed that the system be continued for nine Centres and that a 3 yearly review should be carried out, so that adjustments can be made as to which Centres are recognised according to workload and results obtained at the time of each review, and that funding be allocated based on the findings of the 3 yearly review." The Department of Health considered these proposals and the Secretary of State announced that the neonatal and infant cardiac surgery service would leave the supraregional service arrangements on 31 March 1994. From this announcement, I understand that protective funding was abolished and the regulation left to market forces.

The training of paediatric cardiac surgeons is another topic I would like to discuss in this paper. In my view, we never have had a satisfactory training programme. Trainees have usually been advised to complete training in adult cardiothoracic surgery and then specialise in surgery of congenital heart defects. In practice, this meant 1 year spent in an established department of paediatric cardiac surgery in the UK or abroad. However, this approach had several drawbacks. A 3rd or 4th year senior registrar coming to the department of paediatric cardiac surgery often had no experience in paediatrics or neonatology, in caring for children and their parents, in pain control and drug and fluid requirements of neonates and infants. He had to learn these skills from nurses and his juniors.

The Society of Thoracic and Cardiac Surgeons of Great Britain and Ireland recommended that one trained congenital heart surgeon only was needed each year to cover the needs of England and Wales. The current need may be even less. This fact has to be reconciled with the current regulations that permit the residents to work up to 80 h/week, which means that each department should have four residents. Currently, 12 departments are involved in surgery of congenital heart defects. To resolve these discrepancies, perhaps different programmes for trainee and non-trainee residents will have to be considered.

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<th>Surgeon</th>
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<td>251</td>
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<tr>
<td>Least busy</td>
<td>67</td>
<td>1.6</td>
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It is important that a future consultant paediatric cardiac surgeon should be exposed to as many operations as possible. We do not know how many operations the trainee should perform or assist to become fully trained. The number will differ from trainee to trainee, but I would propose that it should be no less than 300 operations. The quality of training will partly depend on the volume of work that his teacher is involved in. From the data collected by the Working Party, I have estimated that the busiest surgeon in England and Wales performed about 251 procedures per year in 1991, and the least busy surgeon 67. If we consider holidays, study leave and other absences, we may say that the busiest surgeon performs about 6.2 operations per week, compared with the least busy surgeon who only does 1.6 (Table I). The analysis of 571 consecutive infant admissions to our department has shown that truncus arteriosus occurred in only 1% and pulmonary atresia with VSD and major aortopulmonary collaterals in only 0.5% of cases. This means that the trainee working with the least busy surgeon would see only two truncuses and one complex pulmonary atresia in each 3 year period.

I have just described the current situation, but we expect that the number of patients will decrease in the future. Prenatal diagnosis of congenital heart defects has been established in a few specialised centres during recent years. It has been reported that after positive diagnosis many parents opted for termination of pregnancy, rather than for early treatment. The termination rates were 20%, 42% and 61%, respectively, in three departments (London, Paris, Milan) dealing with large numbers of prenatal diagnoses (6). If every pregnancy were screened by an expert investigator, and if the percentage of terminations of pregnancy remained at the levels mentioned above, the number of operations performed in the country would decrease. For example, open heart procedures in infants would decrease from the current 1600 to about 600 per year if the termination rate was 60% (Fig. 3). If there were no reduction in the number of centres and surgeons operating on CHD, the impact on an individual surgeon's practice would be considerable. The busiest surgeon would be performing only 2.5 operations per week and the least busy only 0.7 per week (Table II).

So far, I have discussed achievements in the treatment of congenital heart defects, organisation and teaching. Last, but not least, I would like to look at these achievements from the point of view of patients and their parents. There are several issues. Do we inform the parents adequately about the operation and the future of their child? When we talk about correction of the defect, is it truly correction? I discussed this question in my address...
to the American Association of Thoracic Surgeons a few years ago (8). The definition of 'correction' according to the Oxford Dictionary is "to set right, to substitute right for wrong and to bring into accordance with a good standard". We can therefore consider an operation to be corrective if it achieves normal function, normal life expectancy, without the need for any medication. But do we have such operations? I would suggest that using these criteria we only correct two defects, persistent ductus arteriosus and atrial septal defect. And even these two with the proviso that we operate before pulmonary vascular disease develops.

Why do we not 'correct' other defects such as coarctation of the aorta? It is a simple lesion: we just remove the narrow segment and join the ends together. So why is it not correction? There are several reasons. First, we now know that some patients will remain hypertensive or develop systemic hypertension. Much worse, some patients treated with a particular technique, such as patch aortoplasty, may develop severe complications later. We now have evidence that aneurysm at the site of repair can develop as late as 25 years after the initial operation. Such an aneurysm may perforate suddenly; even elective repair of aneurysm carries considerable risk. Other defects, for example, VSD, Fallot's tetralogy and atrioventricular septal defect, may develop late complications and may require reoperation. This will result in additional stress for the patient and the family and increased mortality.

If we consider occlusion of PDA and closure of ASD as the only corrective operations, what should we call the rest? I believe that there may not be much difference in the final outcome of the operation that is called 'corrective' or 'palliative'. An example is the Rastelli operation which is known as correction, yet the patient may require one or more conduit replacements during his or her lifetime. The patient may accept the need for this '20 000 mile service', but he/she will not have an exactly normal life. On the other hand, there are operations that clearly come under the heading of palliations, and yet give the patient many years of acceptable life without further intervention.

There are, however, other palliations where the outcome is more dubious. The patient with pulmonary atresia, ventricular septal defect and several major aortopulmonary collaterals may require two to three palliative operations, sometimes during the first year or two of life. For some of these, there will be no future. No other surgery could be performed, the quality of life will be poor and often they will die within the next few years. If we could identify such patients, I believe we should advise the parents not to start the series of palliations in the first instance.

The family of a child with serious CHD needs considerable support. It is important that we consider the impact of prolonged hospital stay and repeat operations on the family. In 1966 I joined the Cardiothoracic Unit at Great Ormond Street Hospital as a Senior Registrar. I was impressed by how well the Department was able to get children through a difficult and prolonged postoperative course. At that time, I collected data on eight children who spent more than 6 months on the ventilator, who subsequently recovered and were discharged from the hospital. I reviewed those patients and their families 4 years later. All eight patients were still well, but the parents of six of them had divorced.

Finally, I should come to the title of my article and speculate about future directions. Our aim must be improvement of survival rates and quality of life of our patients. We must strive to achieve results over the whole country that are as close as possible to the results of the best centres. We will need to make some changes in the organisation of care of patients with CHD. We should make provisions for prenatal diagnosis and possibly intrauterine surgery. In addition to cardiac surgery in infants and children, we should provide expert care for adolescents and adults with CHD. We must also treat efficiently newly acquired problems, such as coronary artery disease, valvular disease and arrhythmias, which may be encountered in the population of survivors of early surgery for CHD. To achieve this goal, we shall need different specialists working closely together, ideally under the same roof. I would like to propose the development of centres which will diagnose and treat patients with CHD irrespective of age.

At present, the Ministry of Health has abandoned the principle of Supraregional Centres for infant cardiac surgery. We have evidence that the systems of medical care based on market forces have failed paediatric cardiac surgery in other countries. We know that children with congenital heart defects have been, and still are, operated

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**Figure 3.** Speculation about the decrease in the total number of operations performed for congenital heart defects in infants at various rates of termination of pregnancy.

**Table II.** Speculation about the number of operations performed by the busiest and least busy surgeon after the reduction in number of patients with congenital heart defects by 20%, 40% and 60%

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<th>Busiest</th>
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<td>1991</td>
<td>6.2</td>
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<td>Reduced at 20%</td>
<td>5.0</td>
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<td>40%</td>
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<td>60%</td>
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in some countries by surgeons not well trained in surgery of congenital heart defects with consequent substandard results. There is no justification for this. In order to prevent this happening, we should act now. We should build centres of excellence, whether we call these Supraregional Centres or not. I believe that such centres will be in a better position to overcome the problems associated with the decreasing number of children born with congenital heart defects, both in terms of patient care and training. Such centres will also be well equipped to study and monitor long-term results. In my view, the centres of excellence will eventually provide a better service at a lower cost. When the number of children with congenital heart defects decreases (6), certain complex lesions will be seen exceedingly rarely by individual surgeons. Then, the time may come to operate on such patients in a few national or even international centres.

Declining numbers of children born with congenital heart defects will also have implications for training. We shall need to train even fewer congenital heart surgeons than today. I suggest that the only way forward is to make the training custom-made. Basic training in adult cardiac and thoracic surgery should be retained. In addition, experience in paediatrics, neonatology, paediatric cardiology and research should be included. The actual training in surgery of congenital heart defects should last at least 2 years. I believe I have already made the case that the training should be in a centre dealing with a large number of patients. We will also need to look for new teaching techniques. For example, video tapes can provide an excellent opportunity to teach the details of operative technique. Another technique, which in my opinion will revolutionise surgical teaching, is virtual reality.

We should always keep in mind that the health of our patients matters more than the financial health of hospitals and purchasers. To build an excellent service, the profession should lead the way and not wait for measures to be dictated by the administrators. We must be accountable and monitor our results and make them available to the public. There is no justification for the current secrecy. The results should be judged not by local but by national and international standards.

References

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