The Future of Pediatric Open-Heart Surgery

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George Santayana said that "those who do not study history are forced to relive it." And, therefore, a discussion of the future of pediatric open heart surgery is probably best started by a recollection of what has gone before. Perhaps, particularly, of who has gone before, because people are the cradle of ideas.

Dr. Robert Gross' first successful operation for closure of a patent ductus arteriosus on August 26, 1938, could be thought of as the introduction to modern pediatric cardiac surgery.[1] But even this was preceded by an annotation of Dr. John Munro,[2] also of Boston, published in 1907 which suggested that the patent ductus could, and should, be ligated. But between 1907 and 1938 many bridges of fact and concept had to be built, before a 7½-year-old girl could successfully undergo her operation on that summer day.

A few years later, in October 1944, while Europe was still in the crushing throes of World War II, Dr. Clarence Crafoord resected a coarctation of the aorta in a 12-year-old boy.[3] And these two milestones—ductus ligation and coarct resection—showed that, at least, the aorta could be cross-clamped, opened and resewn without ill effect.

About the same time, Dr. Alfred Blalock at Johns Hopkins culminated his efforts to provide palliation for children with cyanotic heart disease by making a subclavian-pulmonary anastomosis on a small boy.[4]

The story of the origin of the Blalock-Taussig operation must be the classic prototype for all developments in cardiovascular surgery. The observation by Dr. Helen Taussig that children with tetralogy of Fallot deteriorated and became more blue after natural closure of the patent ductus, the concept of surgically constructing a ductus, the fortuitous conjunction of efforts with a surgeon who had already made such an artificial ductus for the experimental production of pulmonary hypertension—all lead to a blaze of surgical effort and success almost unsurpassed.

During the next few years hundreds of cyanosed children were brought to Baltimore, and, across the world, to other centers. Brock, Potts, Dubost, and others strove, not only to emulate, but to enlarge upon Blalock's germinal efforts.

But these operations, while startling in their simplicity and success were yet unable to attack the basic problem. The heart could not yet be opened and repaired directly.

In 1950, Dr. Wilfred Bigelow presented before the American Surgical Association a paper on inflow occlusion to the heart of the dog, and cessation of circulation with survival, all with the aid of hypothermia.[5] Cooling the body reduced metabolic demands enabling circulatory occlusion for 15 minutes at 20°C. Of the 39 dogs operated on by Bigelow, 20 survived rewarming: 6 survived completely. Some of his listeners were not impressed. But others, particularly John Lewis of Minnesota and Henry Swan of Denver

Article available at https://ject.edpsciences.org or https://doi.org/10.1051/ject/197911142

were impressed, not by the number of animals which died, but that any of them survived. The door to success had been opened, but was still only ajar.

In 1952, the door was opened wider by the successful closure by Dr. John Lewis of an atrial septal defect using hypothermia and circulatory arrest under inflow occlusion. A few months later, Dr. Henry Swan, in Denver, repeated Lewis' success and embarked upon a series of operations, experimental studies and reports which signalled to the world that the heart could be safely opened, repaired, and closed.[6]

But this technique permitted no more than 6–8 minutes during which the heart could be opened. Only relatively simple operations could be carried out during this constrained period. The door had been opened further but only to reveal another door still closed.

Dr. John and Mary Gibbon began their research into the development of the heart-lung machine while at Harvard in the mid 1930's. The purpose of their machine whose perfection was delayed by the intervening years of World War II was not, at first, to help in treating heart disease, but was to enable operations on patients with acute massive pulmonary embolism. The apparatus was to provide circulatory and respiratory support while the embolus was removed: to avoid the Trendelenburg operation which Edward Churchill had described as "an early postmortem examination rather than a surgical operation."

In 1953, John Gibbon's work came to fruition with the successful closure of an atrial septal defect in a young woman. True open-heart surgery was now a possibility.[7] It was possible to support circulation and respiration while the heart was opened without having to complete the intracardiac repair in six minutes. Another door was open.

In 1955, Dr. C. Walton Lillehei described his experiences with the repair of tetralogy of Fallot: "total correction." [8] In those cases, circulatory support was provided not by John Gibbon's machine, but by a cross-over circulation from the mother. In discussing Dr. Lillehei's paper, Dr. Blalock correctly foresaw that in the future such operations would be done with the aid of a heart-lung machine, rather than with a biological support system.

There have been a few major landmarks in cardiac surgery which in concept and execution have advanced our ideas in a quantum manner. Dr. Gibbon's success was one such landmark. For, within a few years, almost every congenital cardiac lesion was being attacked—if not always repaired.

Surgery for tetralogy of Fallot serves as an example of the progression of cardiac surgical thought. A palliative operation became available because correction was not possible. With new technology, the heart-lung machine, repair was possible. But for many years, the standard approach was first to do a palliative operation followed by total repair a few years later. Repair at an earlier age would obviously have been desirable, but for lack of knowledge and because of other technical deficiencies it was not possible.

In the late 1960's two developments made early primary repair possible. First was the re-discovery by the Japanese of the usefulness of circulatory arrest under deep hypothermia.[9] But this time the body temperature was reduced to 20°C, not just to 30°C. Prolonged circulatory arrest and, therefore, complicated operations were possible. The second advance was made by Dr. Albert Starr, who, dissatisfied with the poor results of palliative shunts, had begun doing primary reparative operations on children under four years of age with tetralogy. Mortality was lowered and results improved.

Sir Brian Barratt-Boyes in New Zealand,[10] and others, combining profound hypothermia with cardiopulmonary bypass operated on the smallest infants showed that even children under one year of age could be operated upon, and that results were probably better at that age than in older children. Another door had been opened which enabled a vast new group of patients to be operated upon. For, of children born with congenital heart disease, 50% died within the first year of life. It should now be possible to operate on these infants under the perfect conditions of a motionless, bloodless field.

What of the future? There are few lesions, and those mostly rare, which cannot be successfully corrected or substantially palliated. But yet there must be other doors to open. Table 1 shows the signposts to the future.

Better diagnosis. Cardiac catheterization, although accurate, precise and informative is invasive and not without risk in very small, very sick infants. Through the use of newer techniques of echocardiography and radionuclide scanning, easier and quicker, noninvasive methods of diagnosis will be developed. CAT scanning may eventually provide cheap, quick, noninvasive anatomic appraisal of congenital cardiac lesions.

Better decisions. The principle objective of surgery is to change the natural history of disease. But at what point in a natural history curve are the risks of operation warranted. What should be done, why should it be done, and when and how? Knowledge of ventricular septal defects now show that 20% close spontaneously. We know better when to wait and when to operate. Such improvement in decision making will save lives by demanding later or earlier operations—or no operation at all.

Better surgical management must consist of improvements in preoperative, intraoperative and postoperative care. Better decisions about when to operate, and improved preoperative preparation will help results. The extreme importance of pH control, and the avoidance of acidosis and spontaneous hypothermia in neonates have only recently been realized. Circulatory support by balloon pumping or similar means, may eventually become possible, even for small children.

TABLE I

The Future

The Problem	Possible Solutions
I. Better diagnosis	Noninvasive methods Echocardiography Radionuclide scanning CAT scanning
II. Better decisions	Knowledge of natural history: When, why, how to operate?
II. Better surgical management	
(a) Pre-operative Care	Better decisions Better preparation
(b) Intra-operative Care	Perfusion techniques Myocardial preservation New operations
(c) Post-operative Care	Monitoring Long-term support Control of pulmonary hypertension 20–30 year follow-up

Intraoperative care will improve with greater understanding of perfusion physiology and techniques. What is the best way to preserve myocardium? At the moment surgeons sway backwards and forwards like seaweed in an ocean of uncertainty in trying to decide what method to use to preserve myocardium. That patients survive may be more of a tribute to the resistance of the body to iatrogenic trauma than to the precision of science.

At the moment the use of a hypothermic cardioplegic technique seems a reasonable and safe approach. The tenets of this practice are (a) instant arrest to diminish anerobic metabolism, (b) avoidance of intramyocardial edema, (c) balance myocardial pH, (d) stabilize lysosomal membranes with steroids or procaine, (e) provide substrate augmentation with supplemental glucose, and, perhaps, with insulin, (f) and cool the myocardium to diminish oxygen requirements.

Maximum efforts should be made to cool the heart rapidly and keep it cool. Myocardial temperature should be measured, to assure that this goal sought has been achieved.

Postoperative care has improved greatly in past years. Respiratory care, the monitoring of intracardiac pressures, thermodilution measurement of cardiac output all permit a measured, logical approach to cardiac therapy. But the methods are crude and invasive. Better, noninvasive methods will be developed.

And, finally, our system of follow-up care must extend for many years to assure us that we are indeed doing the right thing, to the right patients at the right time.

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