Transcatheter Treatment of Congenital Heart Disease

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IN NORTHERN SPAIN, not far from Altamira with its magnificent polychrome frescoes and Stone Age animals, is the cave of Pindal. Located on a beautiful rocky rugged coast, it is nearly inaccessible and its entrance almost hidden. At first sight the cave is unprepossessing, leaving one unprepared to find what may well be man's first anatomical picture; for it was here that Aurignacian man, some 20,000 years ago, drew the outlines of a mammoth and, in red ochre, marked the heart (fig. 1). Of course, we can only speculate whether he really meant the heart; but, as a hunter, he certainly must have known that to strike the heart was the fastest and easiest way to achieve a kill. He stalked huge animals and was in great danger if his spear thrusts did not score a direct hit. Moreover, he would have to track the animal for many wearying miles or lose it completely. Paleontologists, based on the analysis of many cave drawings, have concluded that the arrows shown on these prehistoric pictographs (fig. 1) were intended to show that striking the heart was the sure way to kill. Thus, it is almost certain that prehistoric man knew of the heart, and probably something of its function.

Illness is older than man. Several dinosaurs unearthed in our Midwest have been discovered to have various types of tumors. One from Kansas had an osteoma (fig. 2A). A microscopic section of that tumor is shown in figure 2B. Man's awareness of illness of the heart dates back to at least the twenty-sixth century B.C. In the Yellow Emperor's Book of Medicine, the following is found:

long slow pulse beats mark its good regulation short empty beats prove its disorderly condition quick pulse with more than six heart beats per one respiratory cycle proves a sickness of the heart a broadly slow pulse signifies a deterioration of the disease.

The Ebers papyrus (sixteenth century B.C.)² contains the statement:

on whatever part of the body a physician might put his fingers — on the neck, on the hands, on the region of the heart, on both arms, on both feet — he encounters the heart everywhere, because its vessels are there for all the limbs; this means that it (the heart) speaks through the vessels of all the limbs.

It is difficult to date man's knowledge of congenital heart disease. Autopsy analysis of congenital heart disease is very new, but one type of congenital cardiac malformation must have been observed by the ancients. A Babylonian tablet (fig. 3), one of hundreds in the British Museum and one of thousands known to be in existence, contains the line: "When a woman gives birth to an infant that has the heart uncovered and that has no skin, the country will suffer from calamities." Thus, the ancient Babylonians, who used abnormal animal and human births for divination, saw and described ectopia cordis. How old is the document? According to Ballantyne,³ writing about this tablet in 1894,

We obtained for the tablet that we have been describing the probable date of 2,000 BC and it can be safely said the observations recorded upon it were much older than this and were possibly a heritage from the ancient Tyrian settlers in the land of the Euphrates. They lived before the time of Sargon I of Agade and through a most extraordinary chance Assyriologists have been able to assign to him the prodigiously ancient date of 3,800 BC.

Cyanosis has long been recognized. Many illustrations of blue-skinned Egyptians may be seen in their necropolis frescoes, and they described cyanosis in the Smith papyrus (c. twenty-sixth century B.C.) (fig. 4). Of course, the Egyptians did not diagnose cyanotic congenital heart disease, but the fate of such a patient was no different from what it would be for the next 4500 years. Only in the last half of the twentieth century has the lot of such a patient improved. For it was then that Drs. Helen Taussig and Alfred Blalock devised the first effective treatment for patients with cyanotic heart disease.

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Dr. Taussig followed her pioneering work on the understanding and treatment of congenital malformations of the heart with the publication in 1947 of her classic book "Congenital Malformations of the Heart." Everyone knows what a classic is: a book everyone talks about but no one reads. I urge my young colleagues to beg, borrow, or steal a copy, for they will find it immensely rewarding. They will also be humbled when reading of the depth of knowledge Dr. Taussig assembled before catheters, before echoes, before scans. I treasure my own two copies of the book, one of which has been inscribed several times by Dr. Taussig. In the other copy, I keep a letter that I received from Dr. Taussig in 1964. That year, the Second National Congress on Cardiology was held in Washington, and I have a vision of Dr. Taussig sitting on the podium, one leg in a full cast, propped up on a stool, chairing the sessions on Pediatric Cardiology for 3 days, morning, afternoon and evening, and having...
available the next morning, a written summary of what had been presented the previous day to be distributed to the attendees at the session. It was at that meeting that I first described some early animal experiments to produce atrial septal defects with catheters. I received a letter of thanks from Dr. Taussig which I cherish and which has served as a continuous source of encouragement and inspiration. It contained the following lines: "It would be wonderful if we can do some of the simpler operations without opening up the chest. I think that is a real advance and a real look into the future."

One of the major advantages of being privileged to deliver this type of lecture is that one may be somewhat anecdotal and speculative rather than requiring that every statement be supported by a mountain of data. I intend to take advantage of that privilege by briefly reviewing our approach at The Children’s Hospital of Philadelphia to therapy of congenital heart disease by cardiac catheter.

Balloon Atrioseptostomy

Our initial attempts, in animals, to produce an atrial septal defect with a catheter device took on several forms. The initial devices were as simple as a bent wire through a large catheter and as complicated as a miniatu- turized bronchoscopic snare. Devices of this sort did work in animals, but were unreliable because sometimes the defect involved the atrial wall rather than the septum. Ultimately, the decision was made to concen- trate on balloon catheters. Early experiments on pup- pies were difficult because the puppy usually has a sealed atrial septum by the time it is big enough for experimentation. The Prince of Serendip must have been at my side when I found a litter of puppies that all had probe-patent foramina ovale. These puppies were subjected to septostomy, and serial autopsies over a 4-month period showed that all the defects remained open. The clinical application is illustrated in figure 5.

In the first 15 years of clinical use at The Children’s Hospital of Philadelphia, we performed over 300 atrio- septostomies. One hundred ninety-two patients had transposition of the great arteries, 31 total anomalous pulmonary venous return, 27 tricuspid atresia, 23 pul- monary atresia and intact ventricular septum, and 28 miscellaneous defects. The results in transposition had a 90% mortality by age 1 year now has a 90% survival rate by that age.

Some of our ancillary observations during the past 15 years are also worth mentioning here. There has been a striking change in the clinical profile of the infant with transposition. In the first 5 years of septostomy, only half of the patients were in the first week of life. In the last five years, almost all have appeared in the first week of life. Indeed, whereas only 13% arrived on the first day of life in the first 5 years, now nearly 70% are seen during the first day of life.

We have also noticed a striking change in the occurrence of subpulmonic stenosis. In the first 5 years, less than 30% of the patients developed this complication. This has increased to an incidence of 62% in the most recent 5-year period. In the last 5 years, 70% of the infants who underwent balloon atrioseptostomy during the first 24 hours of life acquired pulmonic stenosis, many within the first few months after septostomy. Fortunately, our management of these patients is fa- cilitated by the fact that Mustard’s operation can be performed successfully in early infancy.

Figure 4. A hieroglyph from The Smith Papyrus dating from 3000 B.C. Translation (Breasted): ‘‘His lips are cyanotic.’’

Figure 5. Consecutive cineangiographic frames (30 frames/sec) showing a septostomy. Sketches of the frames above, showing the various stages of the procedure, are shown below.
Another observation of interest about transposition was made in our laboratory by Dennis Wood. If one makes a short axial cut across the heart in the normal infant, the cavity of the anterior right ventricle has the shape of a banana and the cavity of the posterior left ventricle has the shape of a grapefruit. In transposition, the situation is reversed, and the anterior right ventricle has the shape of the grapefruit and the posterior left ventricle the shape of a banana. This configuration is well demonstrated on a short-axis, two-dimensional echocardiogram. Figure 6 (top) shows the normal grapefruit-banana relationship of the two ventricles; figure 6 (bottom) shows the reverse situation that occurs in transposition. The width-length ratio of the “banana” affords a reasonable approximation of left ventricular systolic pressure as a percentage of right ventricular systolic pressure. The grapefruit-banana shape relationship is sustained after Mustard’s operation and provides an interesting comment on the biologic adaptability of human tissue.

Transcatheter Closure of Patent Ductus

Our initial studies on ductus closure involved the fabrication of a miniature grappling hook device that could be inserted through the femoral artery of a child, passed into the ductus, extruded from the delivery system and anchored by the hooks into the ductus. The hooks are extremely delicate, and of approximately the same caliber as a 5-0 surgical cardiovascular needle. The prosthesis is anchored to the delivery system by a simple eye pin and sleeve mechanism. It then may be bent upon itself and retracted inside a pod that fits a #6F sheath. In experimenting with this type of device, it became imperative to know the exact location of the ductus on fluoroscopy. In a true lateral view, the pulmonary artery end of the ductus is uniformly related to the anterior edge of the trachea. A properly placed prosthesis must be directly over the trachea or its anterior margin.

Twenty patients were entered into a clinical study of transcatheter closure of patent ductus by the arterial route. Balloon sizing showed that the ductus was too large in three patients. In one of them, two attempts at closure were made, but both prostheses embolized to a peripheral pulmonary artery. Two-year clinical follow-up on this child has failed to reveal any untoward effects. Eight have had successful closure. In nine patients, occlusion was not complete; eight of these patients have had subsequent uneventful surgical closure. In two of these patients, a prosthesis embolized to a peripheral pulmonary artery branch. In each instance, the prosthesis was trapped in a catheter snare and removed without complication. These results, while encouraging, are not adequate. Our experiments had been 100% successful in the calf ductus, but a calf ductus would have closed anyway. It became evident that even a well seated hooked prosthesis could not guarantee complete plugging of the ductus.

The system was redesigned to permit the use of the double-disc, nonhooked prosthesis that could be passed through the pulmonary artery into the aorta and anchored by extruding the first disc in the aorta; pulling it back until it bent on itself in the funnel-shaped ductus; and then pulling the pod off the aftercoming disc until it anchored onto the pulmonary artery side of the ductus. The device (fig. 7) could then be disconnected. This system has been completely tested in animals, and early clinical trials are encouraging. Of eight patients admitted to the study, in three the ductus was too large to attempt, four have been closed completely, and one has had incomplete closure. The patients in the ductus study were 1 month to 13 years old and weighed 2.4–40 kg.

Transcatheter Closure of Atrial Septal Defects

The initial atrial septal defect closure system consisted of a single-disc, hooked prosthesis (collapsed inside a pod) that was delivered across the atrial septal defect, extruded from the pod on the left atrial side of the defect and, with the aid of a centering mechanism,
FIGURE 7. Double-disc patent ductus arteriosus prosthesis. (left) A skeleton in profile. (right) Frontal view of same prosthesis covered by foam disc.

FIGURE 8. Method of implantation of double-disc atrial septal defect prosthesis.

FIGURE 9. Left atrial (A) and right atrial (B) views of cow atrial septal defect, sealed by double-disc atrial septal defect prosthesis.

pulled back to anchor the hooks firmly into the left atrial side of the atrial septum. The prosthesis was released and the carrying mechanism removed.

Twenty-eight patients have been admitted to a clinical atrial septal defect closure study. Closure was not attempted in four cases because the defects were too large (i.e., larger than 25 mm in diameter) and in four others because the defect was considered too small to warrant any risk. Thirteen defects have been adequately closed; in six, closure was not adequate. Five of the latter subsequently had surgical closure, and four were emergencies due to improper implantation. At surgery, removal of the prostheses did not present a problem, and all five surgical procedures resulted in complete closure of the defect. The patients in this study were 14 children and six adults. The children were 4–17 years old.

These results are encouraging, but need improvement. The main problem is that one cannot be certain that tissue will bind to the entire rim of the foam of a single-disc device. Therefore, a double-disc system was produced. Figure 8 is a sketch of the method used. Figure 9 shows the left atrial and right atrial sides of the atrial septum in a cow in which the double-disc device had been in place for 2 months. The defect is sealed on both sides and endothelial covering of both discs is complete. Clinical trials of this system are pending.
In the long run, some atrial septal defects and patent ductus may best be handled with the single-disc, hooked device, and others with the double-disc, hookless one.

I hope that this brief summary of our efforts to treat congenital heart disease without surgery stimulates others to adopt and expand this mode of therapy.

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