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Growth and Development of State of the Art Care for People With Congenital Heart Disease

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For 40 years the American College of Cardiology has been a responsible organization in promoting the art and science of cardiac care for people of all ages. This review chronicles the leapfrogging of medical and surgical creativity and contributions to saving lives and making those lives healthy through informed care for people with congenital heart disease.

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This tale of steady progress honors the American College of Cardiology for contributions during the past 40 years in the art and science of care for patients with congenital heart disease. Our story begins 10 years earlier.

We have done little to prevent congenital heart disease but we have done lots to save lives in the 50 years since Robert Gross successfully tied off a patent ductus arteriosus (1). He did that at the time when diagnostic skills depended on a stethoscope, a three lead electrocardiogram (ECG) and a chest radiograph. Anesthesia was by gas, oxygen and open-drop ether. Recovery took place because of loving care and not with the benefit of antibiotics or even sulfonamides development of cardiac catheterization and uniplane, cut film angiography.

The independent reports by Crafoord and Nylin (3) and Gross and Hufnagel (4) of relief of coarctation of the aorta by resection and end to end anastomosis were soon followed by the method of Brock (5) and Sellers (6) for providing transvalvular relief of valvular pulmonary stenosis by insertion of a valvulotome through an incision in the right ventricle. Closed heart surgery thus became established.

Medical management advanced, too: improved treatment of heart failure by use of cardiac glycosides, rather than digitalis leaf, in dosages based on the weight of the infant or child (7); diuretics were chiefly mercurials. This same period saw successful treatment of bacterial endocarditis with a month of penicillin administration and the promotion of the concept of prevention of that complication through prophylactic use of antibiotics at times of high risk such as after dental extractions. The beneficial effect of 10 days of oral

This article is part of a series of articles celebrating the 40th anniversary of the American College of Cardiology. The series attempts to set the stage for the future by describing current state of the art management of selected major cardiovascular problems and the basic knowledge that will provide directions for advances in diagnosis and therapy.

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penicillin for patients with identified B-hemolytic strep-
lococcal pharyngitis came to be recognized and translated into
programs for primary and secondary prevention of rheu-
matic fever and rheumatic heart disease. Now valvular heart
disease is uncommon.

In this country the American College of Cardiology and
the American Heart Association were leaders not only in the
research and promotion of these programs of treatment and
prevention of rheumatic fever, but also in encouraging the
development of teams of cardiologists and surgeons dedi-
cated to the common goal of saving the lives of people with
congenital heart disease. This became the chief challenge in
heart disease of the young as rheumatic fever began to
decline.

Developments that made possible today's state of the art
care. The 1950s brought diagnosis and treatment to new
heights, with Helen Taussig and Alfred Blalock leading the
way in the team approach in long-term follow-up and training
programs. Diagnostic methodology advanced with bipe
cut film angiocardiology, cineangiocardiology (8) and
the combination of cardiac catheterization with selective
injection of contrast medium, together with techniques for
combined left and right heart studies. Miniaturization of
sampling techniques and development of soft catheters made
it possible to begin to study even the small, sick baby in need
of accurate diagnosis and surgical relief.

Not all handicapping anomalies were extracardiac; ex-
ploration of how to see and repair conditions such as
pulmonary stenosis and atrial septal defect led to the use of
hypothermia and inflow occlusion for treatment of these two
malformations that could be relieved quickly in the limited
time available by this form of circulatory arrest. In Canada,
Bigelow and his coworkers (9) led in the next step forward of
general hypothermia in surgery for congenital heart disease.

Another approach to salvage of babies was for the
common intracardiac anomaly of a large ventricular septal
defect, with failure to thrive and with cardiac failure that
could not be controlled despite intensive medical manage-
ment (10). Surgical palliation (pulmonary artery banding)
was offered by Muller (surgeon) and Dammann (pediatrician)
(11). They created a new lesion, pulmonary stenosis, by
banding the pulmonary artery to stem the excessive flow of
blood through the defect and into the pulmonary artery.

C. Walton Lillehei (12) pioneered open heart surgery as
we know it today with his dramatic use in the mid 1950s, of
controlled cross circulation and then the bubble oxygenator
to close septal defects and repair tetralogy of Fallot. Kirklin
and coworkers (13) adopted the membrane oxygenator of
Gibbons et al. (14) to perfect and expand the art and science
of surgery for congenital heart disease. These two Minnesota
surgeons and their pediatric and medical colleagues were
joined by cardiologists and cardiac surgeons around the
world in quest of definitive repair of as many complex
anomalies as existed in need of repair.

The postoperative complication of postpericardiotomy
syndrome was created when intrapericardial surgery for
heart disease came into being (15,16). This entity required
recognition for what it was and for what it was not (not
sepsis, endocarditis or reactivation of rheumatic fever) and
for the fact that it was self-limited and usually did not recur.
Critical analysis of operative mortality, morbidity and short-
as well as long-term results refined the selection of cases and
improved the results of cardiac surgery.

Interventional pediatric cardiology. The 1960s witnessed
the perfection of catheterization techniques for infants rec-
ognized to be critically in need of medical or surgical help, or
both. Rashkind (17) became the father of interventional
cardiology when he introduced the balloon catheter creation
of an atrial septal defect as life-saving palliation for the
cyanotic newborn with complete transposition of the great
arteries. This accomplishment for such neonates was
matched by the effective surgical physiologic repair in chil-
dren by the venous switch operation of William Mustard
(18). This procedure rerouted venous return to the atria to
match the transposed great arteries, thereby achieving phys-
io logic blood flow and conversion of a sickly, cyanotic
patient to a pink, healthy one.

The 1970s saw two major diagnostic and surgical ad-
vances in this leapfrogging of medical and surgical improve-
ments in the care of congenital heart disease. Let us
consider the surgical accomplishments first. Since the 1950s,
the importance of severe congenital heart disease in young
infants had been appreciated, but the risks in efforts at
surgical salvage were still high. The sad story was that only
half of these critically affected babies reached their first
birthday, and that most deaths due to congenital heart
disease occurred in the first 3 to 6 months of life. A
combination of events in the 1970s changed that: miniatur-
ization of equipment in laboratories and operating rooms,
monitoring and ventilatory support in neonatal and pediatric
intensive care units and skills in sophisticated anesthesio-
logic management. Added to these were the surgical tech-
niques of hypothermia combined with cardiopulmonary by-
pass and the development by Barratt-Boyes and colleagues
(19) in New Zealand of the use of profound hypothermia and
circulatory arrest.

All of these measures accomplished more saving of lives
for babies with cyanotic and acyanotic anomalies. Previ-
ously, such children often died before reaching the suitable
and safe age of 5 or 6 years for surgery with cardiopulmonary
bypass. The door was now open for early repair as the
definitive procedure in infancy rather than later repair in
childhood after palliation in infancy. Babies born with the
common anomalies of large ventricular septal defect or with
tetralogy of Fallot benefited especially by this new approach
to open repair. In this country, Paul Ebert (20) and Aldo
Castaneda (21) and their coworkers led in the excellent
results of surgery for infants with severe congenital heart
disease.

Advances in the diagnosis of congenital heart disease. The
major advance in diagnosis in the 1970s was the advent of
echocardiography, at first M-mode and then two-dimen-
sional (22). At the outset, this imaging technique was validated by comparison with the findings at cardiac catheter-
zation with contrast visualization. As credibility was
dated by comparison with the findings at cardiac catheter-
ization, echocardiography came to be used increasingly
to confirm the clinical diagnosis in mild or moderately severe
conditions where cardiac catheterization would not ordi-
narily be needed, and to be used in serial, noninvasive
follow-up of patients with unoperated or postoperative con-
genital heart disease. Next it began to replace cardiac catheterization as confirmation of diagnosis for patients undergoing cardiac surgery on their malformation (23). This trend increased as Doppler methodology was added in the 1980s.

Recent medical and surgical progress. Other improve-
ments in treatment in the 1970s were both medical and surgical. Management of cardiac failure was helped greatly by the introduction of an effective diuretic, furosemide (24), and by the use, when necessary, of afterload reducing drugs. New antiarrhythmic agents helped to control difficult prob-
lems and new antibiotics were developed that helped cure infections due to resistant organisms.

Pharmacologic manipulations of the ductus arteriosus
often accomplished its closure when it was open and phys-
ologically significant in very small premature babies. The
tagent employed, indomethacin, was a prostaglandin-
synthetase inhibitor (25,26). The converse effect involved
the use of prostaglandin in newborns to maintain temporary
patency of the ductus arteriosus until diagnosis and surgery
could be performed. For instance, a baby whose survival depends on patency of the ductus because of severe congen-
tinal pulmonary atresia or stenosis needs an open ductus to
shunt blood left to right into the pulmonary circuit (27). Alternatively, the infant with severe coarctation of the aorta
also needs a patent ductus to permit a right to left shunt from
the pulmonary artery to the descending aorta to maintain
flow into the descending aorta. The neonate with complete
transposition of the great arteries may benefit from a patent
ductus to provide bidirectional shunting as a temporary
measure. The temporary induction of patency of the ductus
in these critically ill newborns permitted stabilization of their
condition and the avoidance of middle of the night, high risk
cardiac catheterization and emergency surgery.

Surgical inventiveness in the use of external conduits to
connect the right ventricle to the pulmonary artery in con-
ditions of truncus arteriosus and pulmonary atresia was led
by Dwight McGoon and his coworkers (28). Francis Fontan
(29) introduced the revolutionary concept that, in the mal-
formation of tricuspid atresia and rudimentary right ventricle
with diminished pulmonary blood flow, the right atrium
alone could serve to receive systemic venous return and
deliver it to the pulmonary circulation. Fontan, of Bordeaux,
France and William Kreutzer (30), of Buenos Aires, Argentin-
a independently demonstrated that this approach afforded a
new kind of palliation that separated systemic and pulmo-
nary circulations. Jatene and coworkers (31) introduced the
arterial switch operation for transposition of the great arter-
ies. Critical analysis of results modified criteria for selection
for surgery and perioperative management so that mortality
rates began to decline. Surgical suppression of accessory con-
duction pathways began to be used to treat people with
Wolff-Parkinson-White syndrome and serious arrhythmias
retractory to usual drug therapy (52).

The team approach. During the 1960s and 1970s, organ-
ized medicine and the health care system articulated and
formalized guidelines for centers of excellence in the short-
and long-term care of people with congenital heart disease.
The Intersociety Commission on Heart Disease Resources,
headed by cardiologist Irving Wright, published a series of
guidelines for optimal care (33,34). The American College of
Cardiology was a participating society. These guidelines
emphasized the team approach with trained medical and
pediatric cardiologists, cardiac surgeons, anesthesiologists,
pathologists, nurses and related professional personnel and
social services, all working in coordination and cooperative
collaboration in a center well equipped and staffed for
diagnosis and medical-surgical care and follow-up.

The 1980s have seen the flowering of the field of echocar-
diography with Doppler and Doppler color flow mapping
studies that not only image defects but also quantitate
physiologic events of flow and pressure. In many instances
Doppler studies have become the standard, replacing cardiac
catheterization as the first confirmation of diagnosis and in
follow-up. Radionuclide cineangiography (35) and nu-
clear magnetic resonance imaging (36) and digital subtraction
angiography (37) have enhanced imaging capabilities with minimal or no invasive aspect to the study.

Yet the cardiac catheterization laboratories continue to
be busy as they carry out electrophysiologic studies (38) of
extraordinary arrhythmias and perform invasive cardiovas-
cular therapy. Jean Kan and coworkers (39) of Johns Hop-
kins demonstrated the safety and efficacy of balloon dilation
of the moderately or severely stenotic pulmonary valve. This
is now widely accepted as an alternative to open heart
surgery for relief of the obstruction. James Lock and other
interventional cardiologists are carefully performing and
analyzing other balloon dilations that are not yet so success-
ful and remain investigational, such as dilation of native
coaarctation (40), congenital valvular aortic stenosis (41) or
peripheral pulmonary stenosis (42).

Cardiovascular surgical teams proceed with meticulous
care to repair, often in very young infants, even the most
complex of anomalies. William Norwood and colleagues (43)
accepted the ultimate surgical challenge for salvage of a
desperate situation when they carried out the first of several
proposed stages of palliation for the usually lethal and all too
common hypoplastic left heart syndrome. Mechanical valves
are long lasting, and sometimes the native valve may be
rendered functional after surgery. Aortic homografts are
more readily available for external conduits than in the past.
Pacemakers are sophisticated, miniaturized and longer last-
ing (44). Heart transplantation is saving lives, even in
childhood (45), and heart-lung transplantation offers hope of
redemption to some patients with Eisenmenger syndrome.
This recital of developments that took us to where we are
today is liberally laced with surgical names, which pays due
respect to but unfair emphasis on one important group of the
team that brought the care of people with congenital heart
disease to this high state of the art. Procedures tend to have
names attached, whereas medical diagnosis and care do not.
As these surgeons would quickly agree, the success of a
surgical technique depends not only on the concept, the
research and development and the skill in the operating
room. It also requires the correct diagnosis of the patient by
the cardiologist, the careful selection for surgery by both
partners of the team and preoperative and postoperative
team care. Thereafter, their partner in pediatric cardiology
carries out long-term postoperative follow-up with analysis
of outcome so that results can be continually improved.
Many giants in the field of pediatric cardiology contributed
their skills, talents, research, teaching and warm-hearted
care of the patients to this chronicle of the growth and
development of high quality care in congenital heart disease.
Most people with cardiac birth defects do not require cardiac
surgery; they have also been helped by what we have
learned from the surgical experience.

The future. What does the future hold? I wouldn’t dare to
guess! State-of-the-art care as it has developed decade by
decade over the past 50 years has without question saved
lives and made longer lives healthier. It has accomplished
the mission of the American College of Cardiology and
American Heart Association by preventing disability and
premature death for thousands of people born with cardio-
vascular malformations.

Future developments will continue to build on past
knowledge and will employ the team approach with each
member contributing clinical and research abilities and med-
cal, pediatric, surgical, nursing, technical, social, engineer-
ning and laboratory skills in centers with up to date facilities
for ambulatory and in-patient diagnosis and treatment, both
medical and surgical. Early diagnosis and compassionate,
long-term, informed care will always be important. During
the present and on into the future, just as in the past, we do
well to remember that in state of the art care, sympathetic
understanding and knowledgeable care are the key.

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