The official newsletter of the
International Society for Adult Congenital Cardiac Disease

ISACCD update

President's Message

Surgical expertise is reaching new heights. After mastering complex neonatal surgery, our surgical colleagues have transmitted their skills to the new challenges posed by the adult with congenital heart disease (CHD). Multiple previous operations, myocardial (and other organ) dysfunction, chronic cyanosis and arrhythmia are a few such challenges to mention. Furthermore, patient selection and timing and type of surgery remain problematic. Mr. Darryl Shore of the Royal Brompton, keynote speaker at the ISACCD Scientific Session in Orlando last March discussed the challenges of reaching the right decision for patients with aortic valve disease with particular reference to the Ross procedure (see in this issue of our Newsletter).

Dr. Isabelle Vonder-Muhll, Consultant Cardiologist presents the major expansion of the Edmonton ACHD program with a new state-of-the art clinical and research facility being built on site. It is encouraging and welcoming to see new investments being made both in highly trained staff and also on facilities to support clinical and research activity in our field. We should see more of it to catch up with the enormous need for our patients.

I am delighted to report a closer and ever thriving collaboration with adult CHD patient groups. Dr George Warren in his personal and thoughtful contribution to this newsletter as a patient reminds us of the history and the dramatic progress being made in our field. Together with Amy Verstappen, George and the other executive members of the Adult Congenital Heart Association (ACHA) have been a tremendous force raising awareness of the life-long need for tertiary care amongst patients, funding bodies and the public. In close collaboration with ISACCD members -many of whom serve in the Medical Advisory Board of the ACHA- a number of joint ventures are now coming to fruition: the Patient Passport, the Adult CHD Centres' Directory and a fund raising champagne being well on course. We hope to see more of such collaboration ocurring around the world.

Our partnership with the International Journal of Cardiology – our Society's affiliated Journal – has expanded. The Journal's impact factor has increased to the top 30% of the cardiovascular sector. The number and quality of congenital heart manuscripts has also increased. Thank you all for supporting the Journal, for sending in your manuscripts and for offering your precious time for the review process.

Don't miss our forthcoming Scientific Session on Sunday the 13th of November at the AHA Meeting in Dallas. Our Speakers: Dr. Welton M. Gersony, from Columbia-Presbyterian Medical Center, New York, NY "Interventions in Adult Congenital Heart Disease: Evidence-based Decision Making" and Dr. Bob Morrow from Little Rock, AR "ACHD Centers Experience: The Arkansas Children's" promise an excellent session. Be there to support your Society. Bring a senior and/or a junior colleague from your team. Encourage them to join our membership. See you in Dallas!

Michael Gatzoulis

Mission Statement

The purpose of the International Society for Adult Congenital Cardiac Disease is to promote, maintain and pursue excellence in the care of adults with congenital cardiac disease.

The Society is dedicated to the advancement of knowledge and training in medical disciplines pertinent to congenital heart disease in adults.

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International Society for
Adult Congenital Cardiac Disease

When is the Ross Procedure the Treatment of Choice for Aortic Valve Replacement in Adults with Congenital Heart Disease?

Darryl Francis Shore FRCS , Consultant Cardiac Surgeon, & Director, Congenital Cardiac Surgery, Royal Brompton Hospital and National Heart & Lung Institute, London UK

The choice of an aortic valve prosthesis for the adult with congenital heart disease requiring aortic valve replacement demands careful thought. Many of these patients will have undergone previous surgical interventions, particularly previous operations on the left ventricular outflow tract, coarctation repair and closure of ventricular septal defect. Furthermore there may be associated arteriopathy with dilatation or aneurysm of the ascending aorta.

The lifestyle demands of the young adult, anticipated pregnancy, and the need to minimize the number of future procedures all inform the process, prosthesis selection and informed consent. Clinical consideration, including the presence of left ventricular hypertrophy and diastolic or systolic ventricular dysfunction, the need for coronary artery surgery or other intracardiac procedures, and the presence of endocarditis, all influence the choice of prosthesis.

In terms of mechanical valves there are a variety of either single or bileaflet tilting disc valves from which to choose. Mechanical valves offer the most durable alternative, but the risks of anticoagulation and related haemorrhage, and thromboembolism must be fully understood and communicated, and where pregnancy is anticipated the risks associated with anticoagulation must be taken into account. Aitkins suggested a composite linearised rate to compare the performance of mechanical valves and takes into account the incidence of both haemorrhagic and thromboembolic episodes.

Bioprostheses offer the patient freedom from anticoagulation but at the cost of limited durability. The genus of bioprosthetic valves includes stented, bovine or porcine valves, stentless porcine valves, allografts and autografts. In recent years there has been a trend towards the more widespread use of bioprosthesis, partly because the present generation of tissue valves are more durable and it has been widely reported that elective redo aortic valve replacement had a low operative risk. Vogt et al, Sundt et al and O'Brien et al, have reported hospital mortality for elective redo aortic valve replacement, including replacement of homograft valves, of between 0-1.4%.

When compared to stented bioprostheses, stentless bioprostheses have a superior residual pressure gradient and effective orifice areas at rest, and these differences are more pronounced on exercise. They have a greater effect on the regression of left ventricular hypertrophy and improvement on systolic and diastolic function, although as yet there is no direct evidence that this has translated into improved life expectancy. The stentless valve with the longest follow-up is the allograft or homograft valve. It had been hoped that this valve would not exhibit the time-related and the age-related degeneration seen with other types of bioprostheses. This has not proved to be so. Whether the present generation of stentless porcine valves prove superior to allografts in the long run remains to be seen. Durability of bioprostheses is of course crucial in the selection for AVR in young adults.

Enthusiasm for the Ross procedure accelerated when Matsuki et al reported on the longterm results of Ross's pioneering series which demonstrated that degeneration of the pulmonary autograft per se was rarely seen. In a subsequent publication in 1997, Chambers et al showed a 80% survival of the pulmonary autograft at 20 years. The majority of valves explanted showed no degenerative change and the pulmonary autograft regurgitation responsible for their removal was due to technical factors. Between 1988 and 1996 the number of cases reported to the Registry accelerated, however, the initial enthusiasm for the Ross operation has declined largely due to the reported incidence of re-operation required for aortic root dilatation and associated regurgitation, and concern about the fate of the homograft used for pulmonary valve replacement.

David et al reported dilatation of the sinuses and the sinotubular junction in the first 3-4 years after the Ross operation, and that was more common after the Ross operation using the root replace-

continued on page 11

Board Member George Warren

I have joined the ACHA Board this year as a 57 year old congenital heart patient, who happens to be a physician. I was born in Boston in 1947 with Tetralogy of Fallot, had a surgical Potts shunt in 1955, and an open heart "repair" in 1964. While I am of the early generation of surgically treated patients, there were many patients, who were operated on before I was, and I continue to give my thanks to those earlier patients, their courageous families, and the original doctors, nurses and engineers, who gave us our chance to live. I am also mindful of the lives of the animals, mostly dogs, which gave us much needed experimental surgical knowledge, establishing the foundation of early heart surgery.

A trusting family, a wonderful secondary school, and very special, patient friends allowed me to be a normal kid, who got short of breath, rather than a child defined by disability. Like many of us, year by year I had to give up football, hockey, baseball and tennis, but I loved horses, and I was able to play golf successfully. I loved rock and roll, but physically, I couldn't dance to it. Looking at my healthy brother though, the Warren boys were not born to dance. I also accept the fact, that had I been born with the heart of a lion, I still would not have been able to hit a curveball. None of this has kept me from becoming one of the biggest Boston Red Sox fans west of the Mississippi.

I always wanted to be a physician, and I wanted to marry my best friend, Ros, whom I met in my mid-teens. In 1974, Ros and I headed West to Denver, I to begin my training as a pathologist, and she to continue her work in the Federal Bureau of Land Management. For thirty years, I have practiced surgical pathology, with a special interest in diseases of the gastrointestinal tract, and directed my hospital's Chemistry Laboratory. Most of my time has been spent diagnosing surgical specimens and biopsies. Teaching students, Residents, and Gastroenterology Fellows, and assisting some research into the development of cancer of the colon, has provided balance to the daily responsibilities. Five years ago, shortness of breath from my tiring heart returned, and last year I retired.

Being a doctor does not mean I was smart enough to know how to take care of my own heart disease all the time. Going fishing in the Colorado mountains, I passed out at 10,300 feet. One day at the hospital, at age 31, I felt lightheaded and confused, and treated myself with orange juice and cocoa, when I learned the symptoms were due to serious, ventricular tachycardia. When my ankles started getting a little swollen, I treated this sign of early heart failure by using scissors to cut my tight socks a little, before I found my way back to the Adult Congenital Heart program at Boston Children's Hospital.

I have enthusiastically joined the cause of the Adult Congenital Heart Association. Your Board Members are driven to get the word out on a number of issues:

- In many cities, adults with CHD (and sometimes their doctors) do not know enough about the new specialty of Adult Congenital Heart Disease (ACHD) and the measurable peace of mind this specialty provides us.
- We need more clinics specializing in ACHD, and how to locate them through a directory.
- We patients need to know that caring for CHD is a lifelong responsibility, and that we cannot all be "fixed" by a childhood operation.
- We all have to learn to recognize a change in our symptoms.
- We need to carry with us medical bracelets, information of our condition in our wallets, and have computer access to our medical records, should we get into trouble.
- The medical insurance industry needs to admit that disallowing some of us from coverage, or increasing our policy rates 35-40%, because of the way we were born, is unacceptable; the industry refers to profit margins; I refer to the Golden Rule.
- The business community, the charitable private sector, and State and Federal governments need to support clinical care and research programs.

With only two operations, I have had an easier time than many Tetralogy patients. People with a chronic health condition, well managed, know every day what a wonderful world we live in. We are all in it together. Together we need to do some things better. We will pursue these goals, until we attain them.

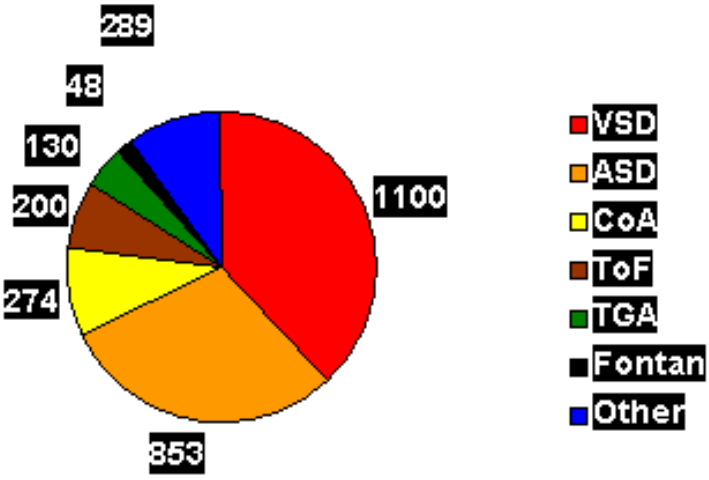
If you are not receiving this newsletter via e-mail, please contact the society at info@isaccd.org or (919) 861-5578, so that we can update our records with your current e-mail address.

The NAACH Program: Serving Adults with Congenital Heart Disease in Northern Alberta, Western Canada and the Canadian North

The Canadian Adult Congenital Heart (CACH) Network was created to pool the knowledge and experience of congenital heart disease professionals in Canada. Created in 1991 and consisting of fifteen centers across Canada, the CACH Network helps strengthen skills and knowledge, and creates a community of individuals committed to caring for adults with congenital heart disease and their families.

The Northern Alberta Adult Congenital Heart (NAACH) program is one of the fifteen CACH Network centers, and is based at the University of Alberta Hospital in Edmonton. As the political capital of the province of Alberta, Edmonton has a referral base of approximately 1 million people. In addition we serve as the surgical referral centre for Winnipeg, Saskatoon, Regina, Calgary and the Canadian north, encompassing an additional population of approximately 1.2 million.

Our program is university based and consists of a collaboration between pediatric cardiology at the Stollery Children's Hospital and adult cardiology at the University of Alberta Hospital. Two adult cardiologists (Dr. Dylan Taylor and Dr. Isabelle Vonder Muhll) and two pediatric cardiologists (Dr. Ruth Collins-Nakai and Dr. Michal Kantoch) provide consultation in the clinic with the assistance of a nurse specialist (Ms. Pam Heggie). We offer a full range of diagnostic and interventional services including ECG, exercise and cardiopulmonary stress testing, echocardiography, cardiac magnetic resonance imaging, cardiac catheterization and electrophysiological testing.



We utilize a database to track congenital heart patients in the NAACH program, and have 2894 patients over the age of 17. The distribution of diagnoses of patients in our program are shown in Figure 1.

Figure 1. Patients in the NAACH program: Diagnoses (numbers of patients)

Not all patients in our database regularly attend clinic. The frequency of clinic visits varies depending on the patient's needs, but most patients are followed on annual or biannual basis. As the population of adults with congenital heart disease grows, our clinic's activity has increased, with a 33% rise in the number of patients seen over the last 4 years (Figure 2).

Figure 2: Growth in NAACH Clinic Activity

In 1998 a percutaneous atrial septal defect (ASD) device closure program was started by Dr. Dylan Taylor. Since its inception, over 205 adult patients have had their ASD successfully closed by device during this single day procedure with a low complication rate. On average, approximately 40

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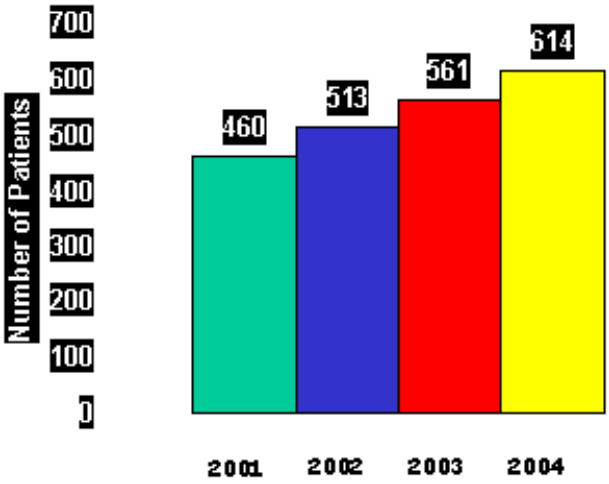

The NAACH Program

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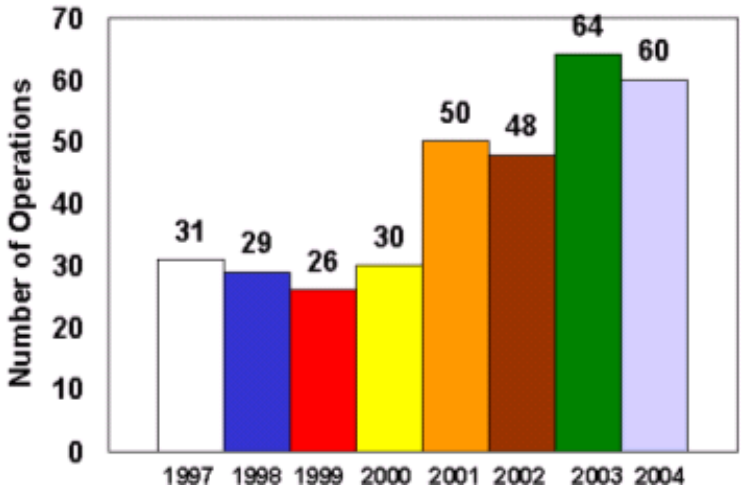
ASD closure procedures are performed per year in the NAACH program.

Our institution serves as a major congenital cardiovascular surgery referral center for our region and Western Canada. Congenital cardiovascular surgery is performed by Dr. Ivan Rebeyka and Dr. David Ross. Like adult congenital clinic visits, adult congenital surgical volume has progressively increased with a doubling in the number of surgical cases between 1997 and 2004 (Figure 3). Currently, we are performing approximately 60 adult congenital heart operations per year.

Figure 3: Growth in NAACH Surgical Activity



Patients attending the NAACH clinic for the first time require a doctor's referral. A typical clinic visit consists of having an ECG, echocardiogram and other tests performed at the time of the clinic visit. Patients first meet the nurse with a focus on patient education, and finally have a consultation with the cardiologist. If necessary, we make further referrals to the Pulmonary Hypertension clinic or Cardiac Transplantation service at the University of Alberta Hospital. Social work, psychology services and genetic counseling are also available if necessary. We are currently in the process of establishing a Maternal Heart Health program for care of women with congenital heart disease in pregnancy.

Certain advantages exist to the patient with adult congenital heart disease (ACHD) in Canada as well as health practitioners working in the ACHD field. These include universal health care which makes diagnostic testing, procedures and consultations available to Canadian citizens without financial penalty. The CACH Network has established a system of regional centers to organize care and concentrate expertise for adult congenital heart patients. Moreover a collaborative predisposition exists between health practitioners to ensure excellence of care for these patients. As an example, we have a weekly teleconference between Edmonton, Calgary, Winnipeg, Saskatoon and Vancouver to discuss case management.

Unique Canadian challenges however also exist which present barriers to the care of adults with congenital heart disease. Our geography means that patients often must travel long distances to access specialized testing and the services of the NAACH clinic. Waiting lists for diagnostic tests, procedures and surgery can delay care. Like many adult congenital heart programs, we struggle with issues related to patients lost to follow-up and the transition of adolescent patients to adult cardiac care.

We are fortunate to have a dedicated team in the NAACH program committed to the ongoing care of adults with congenital heart disease. We look forward to 2007, when the Mazinkowski Alberta Heart Institute will open in Edmonton, a $125 million project to establish a center of excellence for cardiac care in Alberta. This will provide us with further resources and infrastructure for the care of adults with congenital heart disease. For further information, please contact Dr. Isabelle Vonder Muhll at isabellevondermuhll@cha.ab.ca.

ISACCD to Meet in Dallas, Texas!

Sunday, November 13, 2005 • 7:00-9:00 am
Monet Toom, The Wyndham Anatole • 2201 Stemmons Freeway, Dallas, Texas 75207
(214) 746-1200 • www.wyndhamanatole.com

The semi-annual meeting of the International Society for Adult Congenital Cardiac Disease (ISACCD) is scheduled to be held during the Annual Scientific Sessions of the American Heart Association.

The ISACCD meeting will take place Sunday, November 13, 2005, 7:00-9:00 am, at the Wyndham Anatole Hotel.

Lectures will include:

"Interventions in Adult Congenital Heart Disease: Evidence-based Decision Making" – *Welton M. Gersony, MD, Columbia-Presbyterian Medical Center, New York, NY*

"ACHD Centers Experience: The Arkansas Children's Hospital" – *William Robert Morrow, MD, Little Rock, AR*

All are encouraged to attend!
We look forward to seeing you in Dallas!

❑ Yes, I will attend the semi-annual meeting of the ISACCD November 13, 2005, 7:00-9:00 am, in Dallas, TX.

Name _____

Affiliation _____

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Return this form by Friday, November 4, 2005 to:

ISACCD, 1500 Sunday Dr., Suite 102, Raleigh, NC 27607
Fax: (919) 787-4916 • Email: info@isaccd.org






Tentative Program

ISACCD Semi-Annual Meeting in Dallas

Sunday, November 13, 2005 • Monet Room • The Wyndham Anatole • Dallas, TX
Telephone: 214-746-1200 • www.wyndhamanatole.com

7:00-8:30 am **ISACCD General Meeting**

Welcome and Introduction of Speakers
Dr. Gatzoulis

Lecture A: " Interventions in Adult Congenital Heart Disease: Evidence-based Decision Making"
Welton M. Gersony MD, Columbia-Presbyterian Medical Ctr. New York, NY

Lecture B: " ACHD Centers Experience: The Arkansas Children's Hospital"
William Robert Morrow MD, Little Rock, AR

Current membership
Dr. Colman

Treasurers report finances
Dr. Oechslin

International Journal of Cardiology Associate Editor Report
Dr. Gatzoulis

Website report
Dr. Cetta

Newsletter report
Dr. Child

- Should be ready by beginning of Oct 2005 for distribution.
- All contributors to send manuscripts to John Child (Editing) and Peter Kralka (Production) by end of September. MAG will remind contributors. JC to chase up.
- Circulate Newsletter to multiple Email lists (ISACCD, ESC, CACH, Pediheart, AAP, others)

ACHD Centres Database report
Dr. Davidson and Dr. Graham

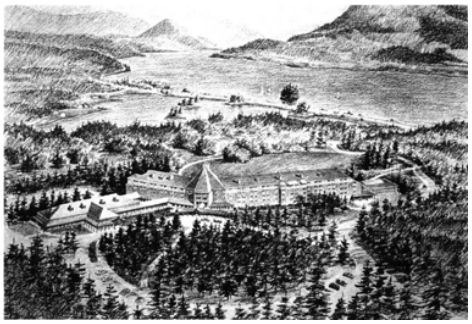
Patients association report
Amy Verstappen

Report from European GUCH Working Group
Dr. Oechslin

Next ISACCD mtg ACC, Orlando March 2006 - suggestions for the scientific program

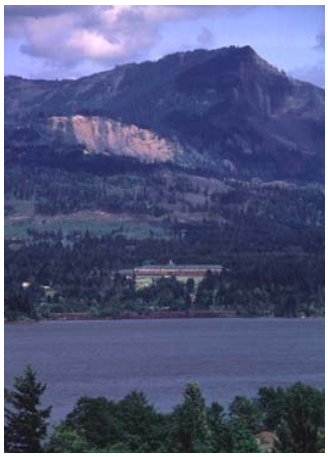
Other business

8:30-9:30 am **ISACCD/ACHA Joint Business Meeting**



16th Annual Congenital Heart Disease in the Adult –
A Combined International Symposium

In Honor of William F. Friedman, MD, FACC (1936 – 2005)



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AMERICAN COLLEGE OF CARDIOLOGY · FOUNDATION ·

5th Advanced Symposium

Congenital Heart Disease in the Adult

Host Institution:
Royal Brompton and the
National Heart &
Lung Institute Programme

Directors:
Dr Michael A Gatzoulis
Mr Darryl F Shore

Registration Information:
CFS Events Ltd
103a High Street, Stevenage
Herts SG1 3HR
Tel: +44 (0)1438 751519
Fax: +44 (0)1438 751520

For more information
www.rbh.nthames.nhs.uk
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25th - 27th September 2006

Venue

Royal College of Surgeons of England
London, England

For further information complete
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5th Advanced Symposium

Congenital Heart Disease in the Adult

25th - 27th September 2006

Venue

Royal College of Surgeons of England

London, England

If you would like further information please complete this reply form and fax it back on the number below, or return it to

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October 2005

Ross Procedure

continued from page 2

ment as opposed to the cylinder inclusion technique. Concern that dilatation of the autograft may be due to the arteriopathy associated with patients with bicuspid aortic valves led Mauro et al to look at the histological abnormalities of the ascending aorta and pulmonary trunk in patients with bicuspid valve disease, and demonstrated that cystic medial necrosis, changes in smooth muscle cell orientation, and elastic fibre fragmentation were more common both in the aorta and more particularly in the pulmonary trunk, in those patients with bicuspid aortic valves.

As most patients with adult congenital heart disease requiring aortic valve replacement have bicuspid aortic valves, it is important to understand whether the presence of a bicuspid pulmonary valve per se has a bearing on the evolution of neo-aortic root dilatation. Several reported series concluded that there was no evidence that bicuspid aortic valve itself was a risk factor for autograft dilatation, neo-aortic regurgitation or re-operation, although some authors have suggested the use of a cylinder inclusion technique for patients with bicuspid aortic valve. Carr-White et al reported no difference between dilatation of the annulus at sinus level or sinotubular junction in patients randomised to either autograft or homograft aortic valve replacement using the aortic root replacement technique in both cases. Furthermore, there was no difference in the incidence of aortic regurgitation in autografts when compared to homografts, and no patient in the autograft group had more than mild aortic regurgitation 4 years following implantation. These authors highlighted the importance of surgical techniques placing emphasis on the removal of all muscle from the homograft, importance of correcting any mismatch between the annulus of the aorta and the pulmonary autograft placing the pulmonary annulus inside the aortic annulus. They also emphasised the importance of performing the distal anastomosis just above the sinotubular junction. Others have emphasised the importance of tailoring or replacing the ascending aorta when there is marked discrepancy between the diameter of the autograft and the ascending aorta.

Carr-White et al also suggested that based on the analysis of stress/strain curves from aortic and pulmonary arterial wall that afterload reduction in the postoperative period would avoid the destruction of collagen fibres in the wall of the autograft, postulated this would allow histological adaptation of the autograft to increased arterial pressure.

The other factor leading to a reduction in enthusiasm for the Ross procedure is the fate of the homograft used for pulmonary valve replacement. It has been reported that there is an increase in the resting gradient across the homograft in the first 3-6 months after insertion and there is a corresponding reduction in effective orifice area. It has been proposed that this is a result of an early inflammatory process and may be reduced by the use of anti-inflammatory medication. Despite these early changes in the long-term follow-up of Ross's pioneering series reported by Chambers et al freedom from reoperation on the pulmonary homograft was 80% at 25 years, when the ethylene-oxide or irradiated homografts were excluded.

It was stated earlier that many adults with congenital heart disease requiring aortic valve replacement have had previous surgical intervention. Some authors have stated that pervious surgery is a contraindication to the Ross procedure. There is no evidence to support this view. However, more time and care is required for excision of the pulmonary trunk which can be particularly difficult where the aortic valve has been previously replaced and where a previously closed VSD encroached on the pulmonary trunk. In a randomised study of pulmonary autograft versus aortic homograft for re-replacement of the aortic valve Carr-White et al reported no difference in morbidity or mortality between the two groups, and at 6 months there was no evidence of any aortic valve dysfunction.

In summary, the Ross procedure should be considered for young adults with adult congenital heart disease requiring aortic valve replacement. It offers a patient superior haemodynamics and valve durability superior to any other form of bioprosthesis. Bicuspid aortic valve, whether or not associated with dilatation of the ascending aorta, previous surgery on the left ventricular outflow tract or closure of VSD, are not contraindications to surgery - however, these features can add to the complexity of surgery. Great attention to technique is required to minimize the risk of postoperative aortic root dilatation and neo-aortic valve regurgitation.