

Anderson's Anatomy of Congenital Heart Disease

There can be no question but that an understanding of the structure of the congenitally malformed heart is the prerequisite for successful diagnosis and surgical treatment. Many centres dealing with paediatric cardiology no longer have access to collections of congenitally malformed hearts, and often, even in those centres where archives do exist, the expertise is not available for proper demonstration of the invaluable autopsy material. With the recent furore concerning retention of organs, it becomes even more necessary to provide a properly constructed visual archive of this priceless material.

Such a venture has now begun. Working with Professor Michael Tynan, a world-renowned expert in the diagnosis and medical treatment of patients with congenital cardiac malformations, the equally respected expert in the structure of the congenitally malformed heart, Professor Robert Anderson, has embarked on the construction of a series of televised demonstrations which, in the fullness of time, will cover the entire spectrum of congenital heart disease.

In the first of his demonstrations, Professor Anderson has concentrated on the structure of the normal heart, since it is axiomatic that knowledge of the abnormal is impossible without a firm grasp of normal structure. With previously dissected hearts from the archives of Royal Brompton Hospital and Great Ormond Street Children's Hospital, Professor Anderson uses this first demonstration as the basis of those which will follow.

The second demonstration, also now available, concerns the entity usually known now as atrioventricular septal defect. Previously described as "endocardial cushion defect", or "atrioventricular canal malformation", Professor Anderson shows that the archetypical feature of the group is a common atrioventricular junction, even when there are two atrioventricular orifices within the junction as in the "ostium primum" defect. He also shows that other lesions, often placed within the group, such as straddling tricuspid valve, or isolated cleft of the mitral valve, are excluded because they possess separate right and left atrioventricular junctions. This second demonstration will form the basis of his anatomical presentation at the forthcoming World Congress in Toronto, so the availability of his material gives an opportunity for those missing this meeting to obtain a flavour of the data presented.

The presentations, made specifically for the purposes of demonstration of the carefully selected pathological material, and photographed in digital format, are available either as videotapes, in either PAL or NTSC format, or as compact discs. The demonstrations last for one hour and seventy-five minutes, respectively.

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ADDENDUM

Several abstracts were inadvertently excluded from assignment to specific poster presentations. They are presented below. We apologize to their Authors and their Institutions.

Robert M. Freedom, MD, FRCPC Co-Chair

P174A

The adult CHD patient in the pediatric CICU Ristuccia E, Elixson EM Children'S Healthcare Of Atlanta

There are approximately 500,000-600,000 adults with CHD in the USA. Advances in technology has provided many of these patients with survival into adulthood. Our 420 bed pediatric multi-campus teaching institution houses an 18 bed CICU that cares for CHD patients from birth into adulthood. Currently there is no Adult CHD program in Atlanta. Between January 1998 and June 2000 our pediatric CICU cared for 1155 cardiovascular surgical patients including 55 adult CHD patients (21%) requiring 59 procedures. These patients had a maximum age of 47 years. Procedures included: Valves (aortic and mitral) 25/55 or 45%, RV outflow (conduits, PA unifocalization) 7/55 or 13%, LV outflow (aortic root replacement, coarctation, Ross, Kono) 7/55 or 13%, Septal defects (ASD,VSD) 6/55 or 11% and others (Rastelli, Fontan, tumor, etc.) 10/55 or 18%. Our ongoing committment to excellence in patient care challanged us to expand our knowledge base to include those areas pertinent to adult patients. We focused on three areas: increasing staff knowledge of the adult patient, optimizing available resources and a focus on family centered care. Conducting a needs assessment of the staff gave us greater insight into the issues of caring for an adult CHD in a pediatric setting. Unit based educational programs geared towards the care of the adult CHD was provided 24/7. With the input from cardiovascular surgeons and intensivist 'Adult CHD Postoperative Standing Orders' were designed and a bedside cart was assembled to house aditional 'adult' supplies. Within our existing facility, rooms were designated for use by the adult patients, providing privacy and promoting a quieter, family centered environment. Awareness of the needs of caring for the adult in a pediatric CICU led us to proactively provide enhanced staff education and increased sensitivity to adult CHD issues, providing excellence in care to both adult and pediatric patients in our CICU.

P174B

Maximal exercise capacity long-term after surgery in patients with fontan circulation

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The Fontan operation has changed the natural history in patients with univentricular hearts. There are however many unanswered questions as; How is quality of life? What is their exercise capacity? Do they react normally during exercise? MATERIAL: In order to study these questions we have reexamined all surviving patients operated at our institution. They were 20 patients, 10 men and 10 women with the ages of 32,0 yrs (14,6-54,2) and 16,9 yrs (11,6-31,2), respectively. The mean follow-up time was 12,6 years. Of the 20 patients 18 were able to perform a maximal exercise test. METHOD: Exercise tests were performed on a bycycle ergometer with 2 submaximal loads of 6 min. each corresponding to 0,5 W/kg and 1,0 W/kg before the maximal one. This never exceeded 1,5 W/kg. Expired air were continuously analysed on the content of O2 and CO2, thus giving the oxygen uptake, pulmonary ventilation, RQ, respiratory rate and heart rate. The patients did perform two tests at two separate days. They all exercised to exhaustion, which was regarded as their maximal level. Further criteria were a RQ > 1,0, VE/VO2 > 30, and a resp.rate > 40. RESULTS: The mean value for VO2max was 1,35 l/min for women and 1,80 1/min for men, corresponding to 25,2 ml/min and kg BW in both groups. Thus their maximal exercise was markedly reduced and below 50% of expected normal values. The mean values for RQ, resp. rate and VE/VO2 were 1,08, 48 b/min and 45, respectively. Maximal achieved heart rates were as a mean 151 beats/min (women 166 and men 136). SUMMARY: Patients with Fontan circulation have a marked reduced maximal exercise capacity and also a marked low "maximal" heart rate. One possible explanation could be a limited William G. Williams, MD, FRCSC Co-Chair

pulmonary blood flow during exercise. However, to prove this hemodynamic studies are needed. The lower "maximal" heart rate has importance when judging exercise capacity from subnormal exercise loads.

P502B

Effects of continuous ambulotory peritoneal dialysis on left ventricular dimensions and systolic functions in children with end-stage renal failure

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To investigate the effects of continuous ambulatory peritoneal dialysis (CAPD) on left ventricular dimensions and systolic functions, we followed up sixteen children with end-stage renal failure with echocardiography for 12 months. The study population consisted of nine girls and seven boys. Their mean age at the entry of the study was 11.3 + 2.2 years (range: 7-14 years). The aetiology of the end-stage renal failure was vesicouretheral reflux in seven, chronic glomerulonephritis in five, pyelonephritis due to urolithiasis in two and unknown in two patients. Echocardiographic studies were performed before the beginning of the CAPD program and 3rd, 6th, 9th and 12th months of CAPD. End-diastolic and end-systolic diameters of the left ventricle, the thickness of the interventricular septum (IVS), and the left ventricular posterior wall (LVPW) were measured by M-mode echocardiography. Ejection fraction (EF), fractional shortening (FS) and left ventricular mass index were calculated according to guidelines of American Society of Echocardiography. At the end of follow-up period (12 months) there was no statistically significant changes in any of the parameters. At the beginning of the study the mean left ventricular end-diastolic diameter was 40.8 mm, end-systolic diameter was 24.9 mm, the thickness of IVS was 8.7 mm, the thickness of LVPW was 7.3 mm, EF was 68 %, FS was 38 % and LVMI was 105.9 gr/m2. At the end of follow-up left ventricular end-diastolic diameter was 41.0 mm, end-diastolic diameter was 24.6 mm, the thickness of IVS was 7.6 mm, the thickness of LVPW was 6.9 mm, EF was 70 %, FS was 38.9 % and the LVMI was 98.8 gr/m2. In conclusion, CAPD seems to be effective for preserving left ventricular mass and systolic functions in children with end-stage renal failure.

P1188A

Life - threatening arrhythmias of wpw syndrome in children Baksiene D. Gurskis V. Anusaitiene V

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Objectives WPW syndrome has the potential for a wide range of varied arrhythmias. Sudden death in children associated with WPW syndrome also has been reported. Aim The aim of this study was to determine group of risk for life - threatening arrythmias in children Methods We examined 50 patients aged 7.8±0.2 years without structural heart disease and with WPW syndrome. An electrophysiological study (EPS) performed in each of them. Various forms and regiments of electrostimulation (programmed inclusive) performed to assess anterograde conduction and refractory characteristics of the accessory AV conections (AAVC). Results In 50 children 52 AAVC which conducted the anterograde direction revealed. The location of AAVC determined in 46 patients: 10 left - sided, 18 right - sided, 11 - anteroseptal, 7 - posteroseptal. Mean anterograde conduction and ERP of AAVC was 227.2±6.3 imp./min. and 248.2±20.34 ms respectively. The shortest AERP (210.8±35.1 ms) determined in anteroseptal AAVC location. Paroxysms of atrial fibrillation (AF) were provoked in 43.8% patients with septal location. We managed to reveal life threatening arrhythmias (severe hemodynamic disorders with short less than 220 ms AER P of AAVC) in 39% of them. In 4 children AF has degenerated into ventricular fibrillation (in 2 with cardiorespiratory arrest). Conclusion The location and property of anterograde conduction over accessory pathways may be related to the genesis of life - threatening arrythmias in children's WPW syndrome.

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