

CONGENITAL CARDIOLOGY TODAY

News and Information for Pediatric and Congenital Cardiovascular Physicians and Surgeons

Volume 5 / Issue 10
October 2007
North American Edition

Spectrum of Multifocal Atrial Tachycardia in Infants

By Sreekanthan Sundararaghavan, MD;
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Keywords

Congestive Heart Failure, Sustained multifocal atrial tachycardia, Amiodarone, Cardiomyopathy

Introduction

Multifocal atrial tachycardia (MAT) is diagnosed when three different p-wave morphologies are seen in the same lead on an electrocardiogram[1,2]. It occurs more commonly in adults with pulmonary disease or COPD [3,4]. Deal et al [5] has noted a lower incidence of atrial tachycardia compared to accessory pathway-mediated tachycardia in infancy and childhood. MAT appears to occur more commonly during infancy than at other ages during childhood[6]. There are limited studies on MAT in childhood which emphasize non-sustained character, absence of depression of ventricular function and complete resolution of tachycardia, though, in infants with structural heart defects, the natural history may be different.

Knowledge about MAT is somewhat speculative. Most of the case reports and retrospective studies from quaternary centers may present a skewed population with inherent bias. Hence, we report a series of patients with MAT managed and treated at one center with different outcomes.

“MAT appears to occur more commonly during infancy than at other ages during childhood[6]. There are limited studies on MAT in childhood which emphasize non-sustained character, absence of depression of ventricular function and complete resolution of tachycardia, though, in infants with structural heart defects, the natural history may be different.”

Case 1

The first case is a two-and-a-half month old female, presenting to her primary care physician's office with vomiting for one month. Change of feeds was made without resolu-

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CONGENITAL CARDIOLOGY TODAY

Editorial and Subscription Offices
16 Cove Rd, Ste. 200
Westerly, RI 02891 USA

www.CongenitalCardiologyToday.com

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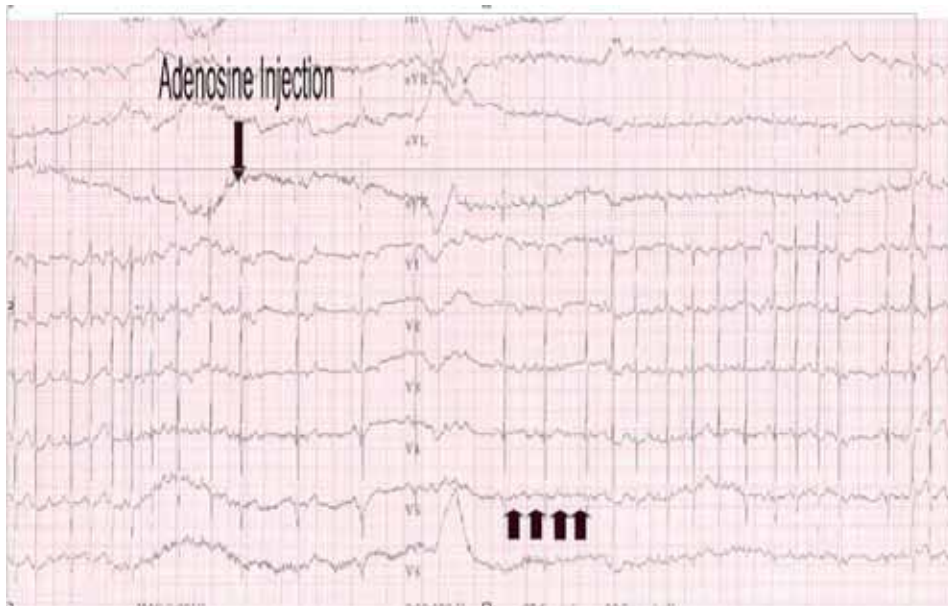


Figure 1. Case 1, Adenosine test reveals atria tachycardia. Small arrows mark P waves, long arrow marks adenosine administration.

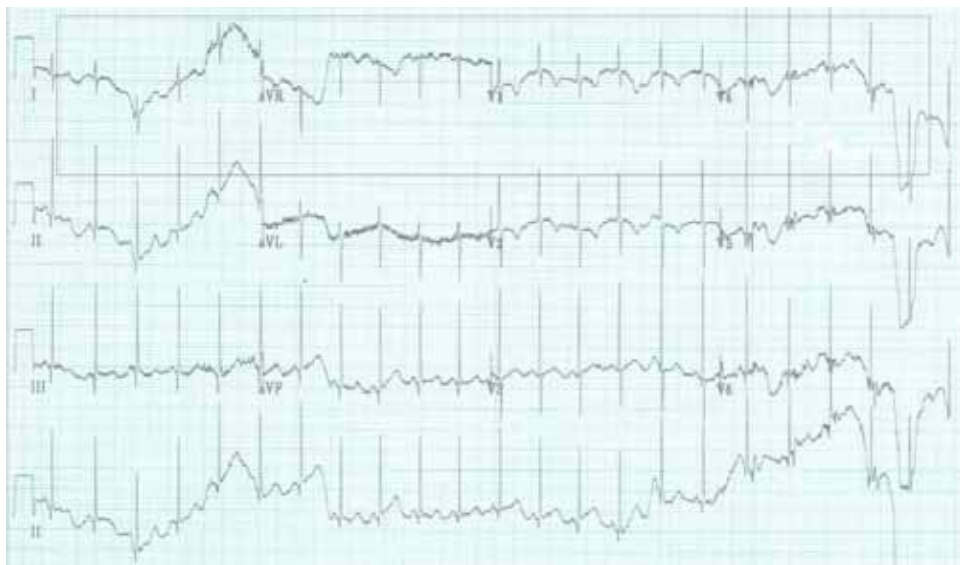


Figure 2. Twelve-lead ECG on Case 1, two months follow-up revealing normal sinus rhythm. Note aberrancy as previously reported by other authors[7].

tion of symptoms. No abnormality of cardiac rhythm was documented during the office visits. She was admitted to the

hospital, for symptoms of upper respiratory tract infection with respiratory distress. On examination, tachycardia (250

beats/minute) with irregular rhythm was found. Twelve-lead ECG demonstrated MAT (Figure 1).

Echocardiogram showed moderately diminished systolic function. Confirmation of the diagnosis of MAT was assisted by administration of adenosine with continuous ECG monitoring (Figure 2). Esmolol was started (100 mcg/kg/min) with rate control (175 beats/minute) and resultant improvement of the distress. The rhythm was irregular despite esmolol dose of 200 mcg/kg/min. She was switched to amiodarone 10mg/kg/day per mouth with withdrawal of esmolol. Follow-up evaluation two months later revealed normal sinus rhythm and normal left ventricular function without pharmacotherapy. The emesis had resolved.

“Knowledge about MAT is somewhat speculative. Most of the case reports and retrospective studies from quaternary centers may present a skewed population with inherent bias.”

Case 2

The second case is a 2 month old male, presenting to his primary care physician for wellcheck, when he was noted to have an irregular heart rhythm although he was asymptomatic. Twelve-lead ECG done in the pediatrician’s office revealed an irregular rhythm. Further evaluation showed a comfortable infant with irregular heart rate of 150 beats/ minute. A 12-lead ECG showed an irregularly irregular rhythm consistent with MAT and associated with an electrocardiogram pattern of right ventricular hypertrophy and left



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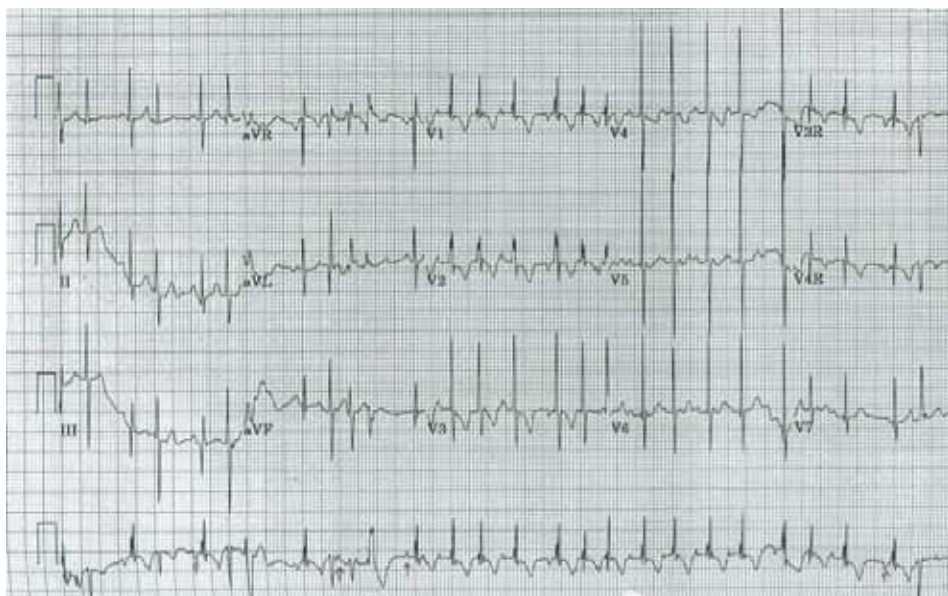


Figure 3. Case 2, Twelve-lead ECG showing Multifocal Atrial Tachycardia (MAT) in structurally normal heart.

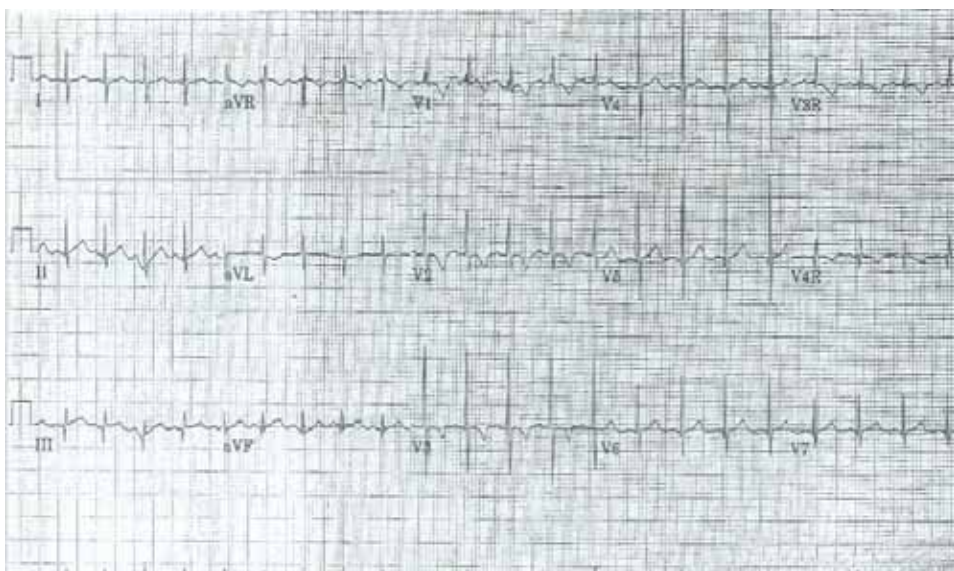




Figure 4. Twelve-lead ECG in Case 2, three weeks follow up revealing normal sinus rhythm.

atrial enlargement (Figure 3). Echocardiogram showed left ventricular dilation and diminished systolic function. A load of Amiodarone (5mg/kg) was given, followed by infusion of 7mcg/kg/min, equivalent to 10mg/kg/day. He remained asymptomatic throughout this time, and two days after admission his heart rate was in the 120's with intermittent irregular rhythm. Amiodarone was changed to oral dosing. By day five, the patient's heart rate was in the low 100's. The

dose of amiodarone was decreased. A follow-up echocardiogram showed normal left ventricular ejection fraction. The patient was discharged home on day seven with heart rate of 95-115 beats/minute. Three months later, amiodarone was discontinued in the setting of normal sinus rhythm on 12-lead ECG (Figure 4). At 4 years, the patient remains asymptomatic, with no evidence of arrhythmia.

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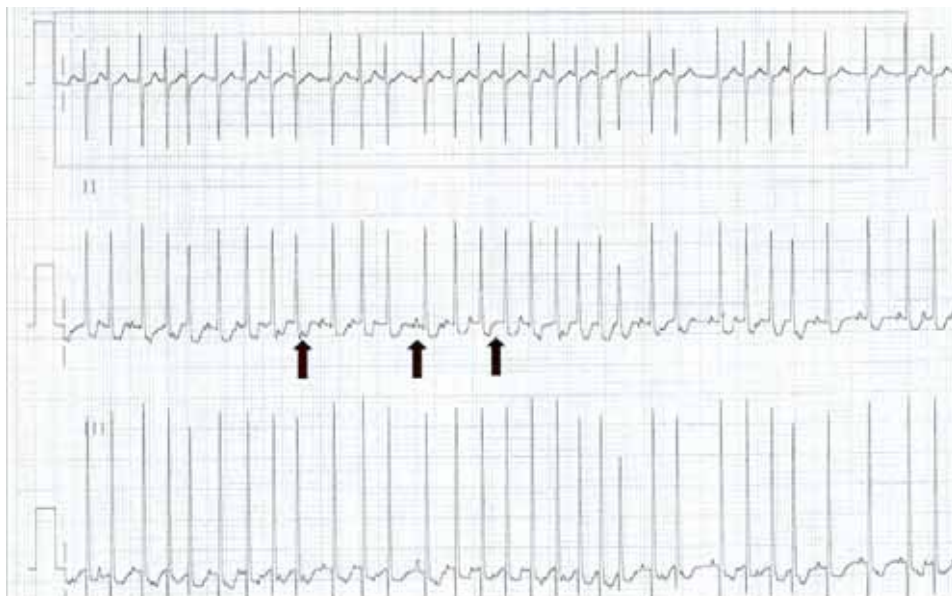


Figure 5. Case 3, MAT in patient with double outlet right ventricle (DORV).

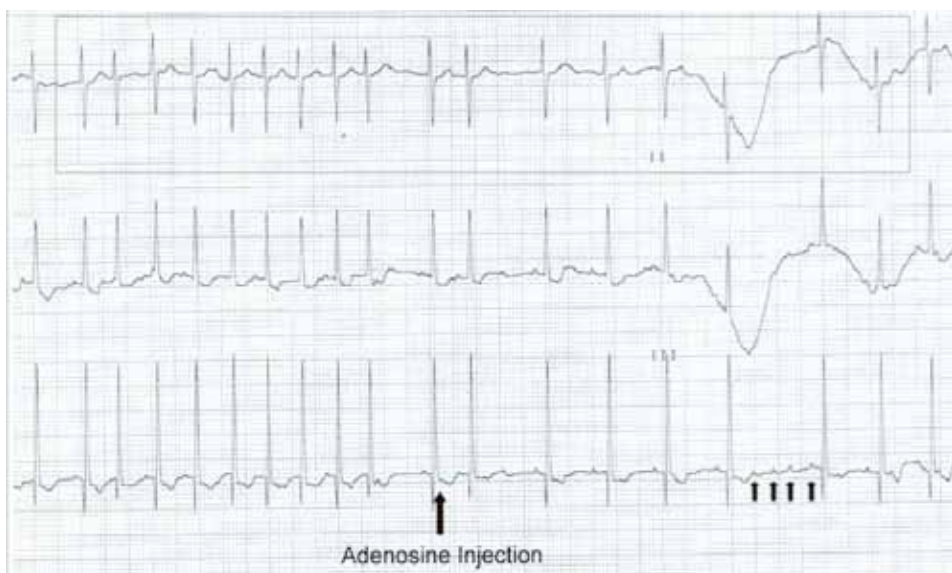


Figure 6. Case 3, Adenosine testing reveals atrial tachycardia. Small arrows mark "P" waves. Long arrow mark adenosine administration.

Case 3

The third case is a 37 week old pre-term neonate born via vaginal delivery. Fetal

ultrasound was diagnostic of double outlet right ventricle with pulmonary stenosis. Pregnancy was complicated by pre-eclampsia and polyhydramnios. At birth,

he had multiple anomalies, including tracheo-esophageal fistula, which was surgically repaired. Later a right modified BT shunt and balloon atrial septostomy were performed. The hospital course was complicated by gram negative bacillary pneumonia and congestive heart failure. At 39 days of age, he developed irregularly irregular rhythm consistent with MAT (Figure 5). Intravenous adenosine was given (100 mcg/kg) to confirm the diagnosis (Figure 6). Subsequently, he remained on mechanical ventilatory support till support was withdrawn on day of life 109 because of the poor prognosis related to the multiple anomalies. During this time, several medications including intravenous esmolol and later intravenous amiodarone, were used and were not effective in controlling the arrhythmia.

Discussion

MAT is an un-familial and somewhat uncommon form of tachycardia during infancy and childhood. The most common forms of SVT are accessory pathway-mediated tachycardia and atrial flutter [5]. In this report, a broad sector of the spectrum of MAT is represented, with findings not previously reported or emphasized by other articles.

Although MAT is a rare form of arrhythmia, our report suggests that the incidence may be higher than previously appreciated. There was a total of 5191 pediatric inpatient hospital admissions from November 1999 to March 2004. During the same period of time, a total of 667 cardiology consultations were obtained with MAT diagnosed in three. Our three cases, thus, present a range from benign to persistent arrhythmia. Cases 1 and 2 are similar in having no structural heart disease, but one of them required multiple medications to achieve benign outcome (Case 2) while the other required a single medication to reach this goal. Conversely, in Case 3, complex congenital heart disease was diagnosed and arrhythmia was resistant to treatment with a rocky post operative period ending with withdrawal of life support.

The initial presentation of MAT could be during a respiratory illness, noticed in 2 of our cases (1 and 2). This association is of extreme importance in general pediatric practice. Careful assessment of the cardiovascular system with minor respiratory illnesses will help in early detection of irregular rhythms in definitive management prior to occurrence of complications, including depression of ventricular function. In one study, Respiratory Syncytial Viral (RSV) infection seemed to have an increased association with MAT[7]. Other studies nearly supported the temporal association with respiratory illnesses[6, 7, 8]. It is intuitive to think of viral myocarditis as the inciting patho-physiology of this type of atrial arrhythmia, but MAT in adults also occurs with chronic respiratory illnesses with evidence of myocarditis, and adult studies have shown an association with COPD[3,4,9,10]. Thus suggests a primary pulmonary-cardiac interaction such as elevations in right sided pressures or alterations in cardiac myocyte properties rather than myocarditis. [1,2,11]

Two patients of our series had episodes severe enough to cause decreased systolic function. This has not been previously reported, probably due to the longer duration of the arrhythmia in contrast with very short episodes reported by others. The overall course, at least in our case series, suggests complete recovery of ventricular function over time with good heart rate control in the interim. This has been supported by other reports on MAT in children[1,2,6,12,13,14,15]. However, the prognosis could be adversely affected in case of associated structural cardiac defects.

The diagnosis of MAT was confirmed in our patients using adenosine administration. This was used in all patients and ruled out atrial flutter with variable conduction and other more rare rhythm disorders. Intravenous adenosine administration is a very useful tool in assessment of MAT. Others have reported wide-complex tachycardia with MAT which was erroneously diagnosed as ventricular tachycardia and treated with lidocaine [7]. This error would be par-

“In summary, MAT should be suspected in pediatric patients with an irregularly irregular tachycardia rhythm having greater than three p-wave morphologies on 12 lead ECG.”

ticularly easy to make in the setting of depressed ventricular function where ventricular tachycardia could be appeared to be likely.

However, with adenosine and attention to clinical findings, the correct diagnosis was made in all of our cases. In the two of our three patients with decreased systolic function at the time of diagnosis, function returned to normal once the tachycardia was controlled. Duration of symptoms prior to diagnosis in our patients was similar to that of patients reported by Bradley et al who had suggested a median duration of 4.9 months and a mean duration of 6.7 months (8-18.5 months) [6]. In Case 1, the tachycardia completely resolved by 2 months, and by 2 weeks in Case 2. We found sustained arrhythmia in all our patients comparable to other authors.

In summary, MAT should be suspected in pediatric patients with an irregularly irregular tachycardia rhythm having greater than three p-wave morphologies on 12 lead ECG. Its spectrum includes sustained arrhythmia and congestive heart failure reversible on successful therapy. Multiple medical therapies may be required to gain control while patients without structural heart disease and MAT will see spontaneous resolution over time.

Acknowledgments

The authors thank Frank Zimmerman, MD at the University of Chicago for his expert input in this article.

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SPENCER TECHNOLOGIES

Interview with Richard Van Praagh - Part 1: Academic Career

By Bradley W. Robinson, MD

"This is the first in a series of two articles drawn from an interview by Dr. Bradley W. Robinson with a pioneer in the field of pediatric cardiology, Richard Van Praagh. In the next few months we will publish the second and final part of the interview, "Family Life."



Richard Van Praagh, graduation picture, June, 1954, Faculty of Medicine, University of Toronto, Ontario, Canada.

Q: How did you develop such an interest for cardiovascular pathology?

It has been really a tremendous, a great joy and a high privilege to be able to take part in this marvelous field. I am a native Canadian and a pediatric cardiologist very much interested in pathology and embryology and molecular genetics. We were encouraged to do a year of basic science in training. I said to my chief resident, Dr. Jim Boone, who was a wonderful man. "What sort of basic science should I do?" Everybody in our clinic thought that maybe pathology was the most clinically relevant of the basic sciences, so I said, "Fine, where should I do it?" The wisdom at that time was that if you could afford to be paid nothing at all, go to work with Sidney

Farber in Boston. But if you had to be paid just a little something, go to work with Dorothy Anderson at Baby's Hospital in New York. I was poor as a church mouse, unmarried of course, because we were paid very, very little. I did my year of pathology at Children's Hospital Boston in Sydney Farber's department under the direction of Dr. John Craig. I discovered several forms of tricuspid atresia that had never been reported, that were previously unknown. Now this was 1956-57, and this was how I became interested in pediatric cardiology.

Next I was a senior assistant resident in pediatrics for Dr. Charles A. Janeway at Children's Hospital Boston. One day he called me into his office and said, "Dick, sit down." I said, "Yes sir!" I was wondering what in the world have I done? He asked me to investigate our patients who were undergoing exchange transfusion because far too many children with erythroblastosis fetalis were dying during and shortly following the exchange transfusion for Rh problems. As you probably know, Lou Diamond and Fred Allen had developed the exchange transfusion at the Boston Children's Hospital. We discovered that there were a lot of things that we could do to improve exchange transfusion. For example, patients were often receiving old, cold, banked blood—weeks old, very cold. The potassium was often very high. The pH was very low. So one of the first things we did was to say, "What about using fresh heparinized warm blood". This made a big difference.

Then I started a study of the causes of death in erythroblastosis fetalis. In the meantime, our group here in Boston had invited Emmett Holt (who thought exchange transfusion was unnecessary) and his crew to come up to Boston from Babies' Hospital in New York. We essentially said, "Okay, here are our Rh babies. Let's do an experiment just to see how his treatment with intravenous diglucuronide works." Then somebody said, "Well, in order to make this a proper scientific experiment, we have to have a control group, right? We'll give these babies intravenous diglucuronide and we'll give those ones over there just 5% dextrose and water basically nothing.

Q: A placebo?

Yes. Then, to everybody's astonishment, the bilirubin curves descended exactly in parallel in both groups and everybody realized immediately that the effect of diglucuronide was just dilutional. So I got introduced early to the urgency and the excitement of research, you see.

Q: You liked cutting edge research?

Yes! We are talking about love. I thought research was unquestionably worthwhile. So then I then did my first year of clinical pediatric cardiology with Dr. Helen Taussig at Johns Hopkins Hospital. I got to know Dr. Alfred Blalock very well. Frank Spencer was there at that time so was Henry Bahnson. Then, in order to get properly trained as a pediatric cardiologist, I knew that I had to get good training in cardiac catheterization and angiocardiology, and just at that time at the Mayo Clinic in Rochester, Minnesota, a brand new lab had been completed and it was going to be run by Dr. Jeremy Swan. I applied and was very fortunate to be accepted. So the next year, I did nothing but cath and angio all day in the Cath Lab.

This is what I had always wanted to do. I had been dragooned into hematology research. I found it fascinating. You see one of the problems with me is I like everything. The reason I chose pediatrics was that it appealed to my gambling blood. In other words, it is not a question of a four or a five-year survival against inexorable forces and age and decay, of slow death. It is a question, if you can do it, of forty or fifty or sixty years' survival, a mother, a father, a future family. I thought that was terrific, so that is why pediatrics.

Q: Did you do research at Mayo?

Let's see, in 1959 and 1960, my project was to figure out single ventricle. You see, single ventricle can mimic all kinds of other conditions that have two ventricles, and what people were discovering was that if a patient with only one ventricle got sent to the Operating Room thinking there were two, and if the surgeon incised the only ventricle that the patient had, then the



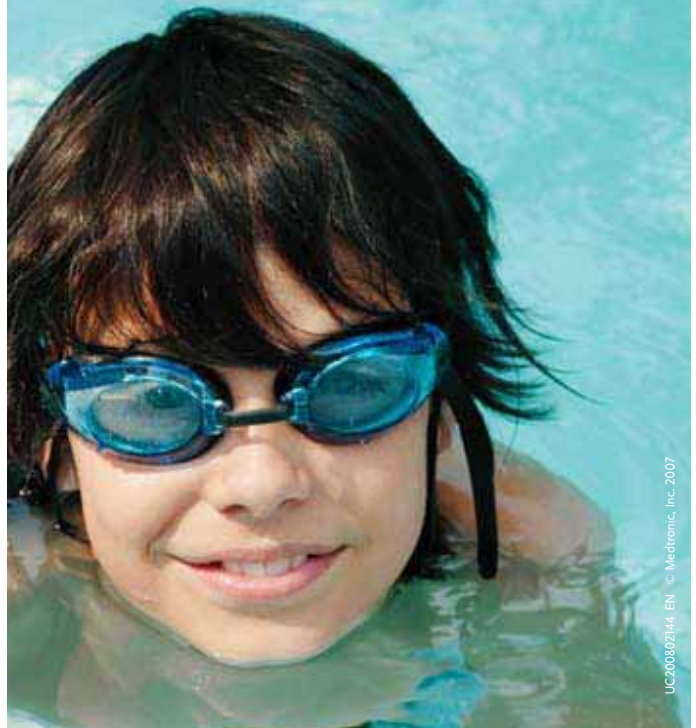
At the graduation banquet of the class of 1954, Faculty of Medicine, University of Toronto (from left to right): Sidney Smith, president of the university; Richard Van Praagh, president of the graduating class; Sir Robert Watson – Watt, the inventor of radar and our guest speaker; and J.A. MacFarland, Dean of the Faculty of Medicine.

essentially inevitable result at that time was death of the patient. So, the premium on understanding single ventricle was very high indeed. So, I worked all day in the Cath Lab and all night in Dr. Jesse Edwards' pathology lab. John Craig, my immediate teacher here at Children's, had told me about the work of a pathologist by the name of Dr. Maurice Lev of Chicago, who had introduced the morphologic approach to the understanding of the various cardiac chambers. So I used an extension of Dr. Lev's approach that didn't depend only on the morphology of the septal surfaces, both of the atria and the ventricles, to identify the morphologically right atrium, right ventricle, left atrium and left ventricle. I realized that the free walls of both ventricles were just as reliable as the septum. Using this understanding, I was able to figure out what single ventricle is. Previously, people had thought that the single ventricle was basically a huge VSD. What we found was that single ventricle is really absence of one or the other ventricle. The commonest one is absence of the right ventricular sinus or body or inflow tract, and much less frequent is absence of the morphologically left ventricle. Then my colleagues said, "Well look, you have to present this to the American Heart Association." So I said, "Fine." But then, various people thought that they should be making the presentation for me because I was very junior. But then I said, "Very probably, you could make a better presentation than I could. But what are you going to do when Dr. Lev stands up, or Dr. Helen Taussig stands up and questions the basic concept? Do you think you'll be able to defend it?" This was the reason that I was allowed to make the presentation. I was told, "Dick, this is



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The founders of the Cardiac Registry – the cardiac pathology lab – of Children’s Hospital Boston, CA 1970: Richard Van Praagh (left) and Dr. Stella Van Praagh (center), with Dr. Masahiko Ando (right) – our first Japanese fellow who subsequently established an outstanding cardiac registry in Tokyo.

the first time that the Mayo Clinic has ever been represented at the national level by a fellow.” So I bit my lip and said nothing. Perhaps I should add that my presentation to the American Heart Association went well. And both Dr. Lev and Dr. Taussig did make questioning comments.

Q: What happened next?

My dear father died, so I went back to Toronto and tried to shore up the family. I was a senior research fellow in pediatric cardiology in the department of Dr. John Keith, who was the Canadian father of pediatric cardiology. I was doing clinical work and research.

Q: Where did you meet Stella?

That’s quite a story. As you may know, dear Stelly died on June the 3, 2006. [EDITOR’S NOTE: *Congenital Cardiology Today* published an article on the life of Dr. Stella Van Praagh entitled, “The World Loses a Gifted Pediatric Cardiologist, Pathologist and Philosopher” in the July 2006 issue. See the past issues section on the website to read this article - www.CongenitalCardiologyToday.com]. We were a team; ours was an affair of the heart - an *affaire du coeur* - in every sense. You see, Stella was my first student; we actually met at Hopkins. I was at one of Dr. Taussig’s reunions. All of Dr. Taussig’s fellows had to make a 10-minute speech on what they

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Director of Echocardiography for the Center: We are seeking board certified physician candidates with experience in advanced echocardiography. Additional experience in other non-invasive imaging modalities would be advantageous to the candidate’s application.

Director of Pediatric Heart Transplant: We are seeking a pediatric cardiologist with a strong clinical background in heart transplant and heart failure.

Pediatric Cardiologist: Board eligible/certified in pediatric cardiology with interest in outpatient and inpatient aspects of their field, including attending satellite clinics in the Chicago metro area. For this position recent fellowship graduates are encouraged to apply.

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were doing. My talk was on the angiocardigraphic appearances of the right ventricular outflow tract in tetralogy of Fallot. After I finished my talk, this charming young lady (Stella) with enormous brown eyes came up and started asking me very, very intelligent questions.

Q: How did you later re-meet her?

Dr. Keith asked me to figure out dextrocardia for a meeting of the Ontario Heart Foundation that was only 10 days away. How many kinds are there, how can we diagnose them, and what can we do about them? Dr. Keith suggested that we combine our data from the Hospital for Sick Children in Toronto with Buffalo Children's Hospital data. Stella had moved there. So, Stella and I started working on dextrocardia. She later told me that she had never worked harder in her life because she had full clinical duties, and then there was me with all of these hearts. So I was then telling Stella that, there is a way of looking at this that greatly simplifies everything. If you analyze these hearts in a segmental way, segment by segment, because all hearts are made up of the three major segments that are the viscera and atria, and then the ventricular loop, and then the conotruncus. If one can diagnose the anatomic type of visceratrial situs, the type of ventricular loop, and the type of conotruncus, also being very careful to note the status of the connecting segments at the atrioventricular junction and the ventriculoarterial, then the whole thing becomes very easy. You don't need to remember any classifications. So, dear Stella was my first student in that sense. And in recompense for that, she was teaching me for the rest of our married lives. She was wonderful. That is how dear Stella and I got to know each other. Then when we announced our engagement to be married, everybody in Toronto said, "Oh, so that's what you were doing!" We had to publish the paper to prove that that wasn't all that we had been doing.

Q: So that was in 1962?

1962 is when we got married. We had three children in three years because neither of us was terribly young. Everybody was beautiful and acyanotic and everything worked out nicely.

Q: In 1965 when Dr. Nadas and Farber and Gross invited you to return, how did you come up with the idea of a pathology registry?

Dr. Alex Nadas, who you know is the American father of pediatric cardiology, was very important. So, too, was Sidney Farber the father of chemotherapy and the Chief of Pathology. Sidney Farber looked at me and said, "Dick, I want you to make this the best lab of its type in the world." So I said, "I'll do my best, sir." We very seldom saw him because he was involved in leukemia research at the Jimmy Fund Building. As a pediatric cardiologist, for me every autopsy was a CPC (clinical pathology conference). I had been to the clinical conferences, you see. If there were any diagnostic mistakes, it was as much my fault as anybody else's. If there was any management thing that might have been done better, again, if I didn't speak up or suggest it, it was as much my fault as anybody else's. So what we would then do, we would sit down with the cardiology fellows and we would go through the chart and I would get the fellows to tell me exactly what the diagnosis was, what are all the lessons of the case, before we started the dissection, because everything is obvious in retrospect. So this was the most wonderful learning opportunity in the hospital. In fact, it was the only place that you could find out if you were right or not, you see. And so it was a wonderful way of learning. This was part of the Cardiology training program and part of the Surgical residency training program, and the radiologists would come, and even the neonatologists. Then, we also welcomed in the cath lab techs, the echo techs, the nurses...



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The Van Praagh family at Richard and Stella's retirement party in June, 2001 (left to right): Alexander, Richard, Stella, Helen and Andrew.

Q: Were you one of the clinicians involved in patient care?

I didn't actually look after the patients directly, but I was the one who was involved in teaching the radiologists how to interpret the angiograms. I was not an academic bystander looking on with academic interest. Every case for me was my patient, a matter of life and death. Then people started coming from all over the world and so we trained people from Japan, from Canada, from all over the States, from France, Germany, Italy, Greece, and England.

Q: Did you pattern the registry after a model or create it as you went along?

Well, I realized what an enormous privilege it was to be a clinician, a cardiologist, you see, because you have to have ques-

tions that need to be answered. I should have told you that in going over each case, we would go through the whole chart, all the cath data, ECGs if present, and then above all, the angios. We would go and look very carefully at everything. I would get the guys to say, okay, what is the diagnosis? What is the problem here? Was the operation perfect, or was it imperfect in some way or incomplete? What could we have done better? Then only after that would we do the dissection and then the lessons would scream at you. We would often say to ourselves, this is unknown. We have to write this up.

And I would get correspondence from, well for just one example, a friend from Halifax called me up and said, "We've got a very weird case (in a live patient). May we send you all the information?" I said, "Of course." So they sent me everything including the angios, and I looked and realized that this was a newly discovered

form of tetralogy of Fallot. The atria were normally located in situs solitus. There was an ordinary D-loop at the ventricular level, but the infundibulum and great arteries were in mirror image. They were inverted.

Q: So you received consultations on live patients such as a patient with tetralogy of Fallot with inverted great vessels?

Exactly. It was a tetralogy of Fallot {S,D,I}. Then, I tried to explain all this over the phone to the surgeon and then the question was, "Well, what are we going to do about it?" I said, "Well, there's one thing you've got to be very careful of and it is the right coronary artery because the right coronary artery always runs right over the infundibulum, exactly where you want to open up, because the aorta is to the left, you see, and then the right coronary runs across the infundibulum. The pulmonary artery is now to the patient's right. So the right coronary artery either needs to be jumped with a conduit or a homograft, or perhaps it could be scived (resected) out beneath to open up an adequate pulmonary outflow tract. You will be able to tell in surgery whichever it is, but be very careful of the right coronary artery." And he was, and the patient did beautifully.

Q: What extracurricular activities did you do growing up?

I was on the high school gym team. Scouting was fun. I love camping and canoeing, canoe tripping. I love skiing. I have enjoyed team sports and tennis. I love debating, which is sort of team and individual. I played on the high school baseball team.

Q: What would you discuss at the dinner table growing up?

Everything and anything. We were intensely interested in international affairs. There would be a lot of discussion about everything, current events, the war, whatever was happening. I kept a scrapbook, which is fascinating, all during the war. We loved (French and English) literature. My brother, Ian, and I went on visites interprovinciales it is called, 'interprovincial visits', to Quebec and lived with a French-speaking family for two weeks where we were not allowed to speak a word of English. We were interested in ... thought in

all of its forms. Nothing was off limits. We had a very privileged background. We studied music, piano and music theory, right up to the teaching level.

Q: How did your summer experience working on the road crew help you with pressure?

We had to become relatively accomplished street fighters just to survive. We never picked fights but it was necessary to learn how to defend oneself in the event. This was extremely helpful later on when people in medical training would occasionally try to pressure you into doing something that you knew wasn't quite right. What I would do, I would just close my eyes and remember back to the road crew and all the fights that we had and then I'd smile. These later academic coercive situations, which I guess all of us get into occasionally, struck me as faintly absurd. My only hope, my only prayer was that I would never lose my temper with anybody, because that would have been extraordinarily counterproductive, as you can imagine. And I never did.

Q: What are you doing now?

I am trying to get a book done. The book is a data-base study of 3400 autopsy cases of heart disease in infants and children, almost all congenital, about 95% congenital and 5% acquired. You see, I retired in 2002 in order to get some work done! This is what I am doing.

I only go to meetings that I can't honorably escape from. When dear old friends call you up and ask you, you know, plead with you, "Please come!" it is very hard to say no. So just to give you an idea of what has been going on recently in my little world, two weeks before my dear wife died, I was in Moscow to give the Bakoulev lecture at the 50th anniversary of the Bakoulev Institute of Cardiovascular Surgery of the Russian Academy of Medical Sciences in Moscow. This was an enormous honor. Then, I went to Tokyo because Atsuyoshi Takao had died. He was the head of the Tokyo Women's Medical College, the Heart Institute of Japan. I was the only Westerner there.

Q: What books on your night stand?

Well you know I am very interested in, as I'm sure we all are, as biologists, "The Journey of Man" by Spencer Wells. Then there is another one called "Before the Dawn," by Nicholas Wade, which is again the early history of homo sapiens. But what I am really reading right now is a mar-

velous book. It is "The Story of Civilization" by Will Durant, who wrote an 11 volume account of the story of civilization. I am re-studying my Latin book because I love language. You know, all of the words we use have a story -- there is a story for each and every one. The etymology is fascinating. What I am really doing of course is studying English, but it is fun.

Q: Do you have any advice you would give to young pediatric cardiologists or young surgeons?


I think that one should get as well trained as possible, and that one should adhere to the truth. For us, as scientists, this is the only thing that matters. Everything else is conversation. Getting well trained at the best places by the best people and trying to do literally the very best that one possibly can in all situations. It is the old Boy Scout motto, "Do your best." It really is, and it comes to that. Not just in every case, but also in your training, and in each and every paper. You asked me what I think is our best paper, and I can't answer that. So I would say to people, just do the very best that you possibly can. in everything. In other words, for me, science is not an occupation. It is a way of looking at everything. It is a way of thought. It is a way of life.

Thank you. This has been a wonderful experience.

CCT



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Congenital Heart Center at the University of Florida

The Congenital Heart Center at the University of Florida, is recruiting a Board Certified Pediatric Cardiologist for the faculty position of Clinical Assistant Professor with experience in echocardiography. This faculty position will be focused in the area of congenital heart disease serving as a general pediatric cardiologist and being primarily responsible for performing echocardiography services. This role includes opportunities to participate in research and teaching efforts of residents, fellows, medical students and other health care professionals.

The appointment will be non-tenure-accruing. This position will remain open until an appropriate candidate is selected

Applicants should send a letter of application, a C.V., and three letters of recommendation referencing LP#00017673 to:

Barry J. Byrne, M.D., Ph.D.
The Congenital Heart Center
University of Florida
College of Medicine
P.O. Box 100296
Gainesville, FL 32610-0296

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Faculty Member for the Congenital Heart Center at the University of Florida

The Congenital Heart Center at the University of Florida is recruiting a Board Certified Pediatric Cardiologist with fourth year interventional catheterization training for faculty position of Pediatric Interventional Cardiologist. This position will assist with coordinating all aspects of pediatric interventional cardiac catheterization services and provide general pediatric cardiology care. This role includes teaching of residents, fellows, medical students and other health care professionals and participation in a strong clinical research program and excellent clinical practice.

The appointment will be at the non-tenure accruing level of Clinical Assistant/Associate Professor based upon experience. This position will remain open until an appropriate candidate is selected.

Applicants should send letter of application, C.V., and three letters of recommendation referencing LP# 00023005 to:

Randal M. Bryant, M.D.
Search Committee Chair
Congenital Heart Center
University of Florida
College of Medicine
P.O. Box 100296
Gainesville, FL 32610-0296

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Faculty Member for the Congenital Heart Center at the University of Florida

The Congenital Heart Center at the University of Florida is recruiting a Board Certified Pediatric Cardiologist for the faculty position of Director, Non-invasive Imaging. This position will lead a cohesive unit for advancing research and clinical care in congenital heart disease imaging through coordination of echocardiography, MRI and CT angiography. The position will also provide general pediatric cardiology services and includes teaching residents, fellows, medical students and other health care professionals.

The appointment will be at the non-tenure or tenure accruing level of Associate Professor or Professor based upon experience. This position will remain open until an appropriate candidate is selected.

Applicants should send letter of application, C.V., and three letters of recommendation referencing LP# 00023002 to:

Barry J. Byrne, M.D., Ph.D.
Medical Director
Congenital Heart Center
University of Florida
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Medical News, Products and Information

AGA Medical Corporation Receives Approval for the AMPLATZER Muscular Ventricular Septal Defect Occluder



AMPLATZER[®] Muscular VSD Occluder
© AGA Medical Corporation

MINNEAPOLIS - (September 10, 2007) - AGA Medical Corporation ("AGA") announced that it has received U.S. Food and Drug Administration (FDA) approval to market the AMPLATZER Muscular Ventricular Septal (VSD) Occluder.

The AMPLATZER Muscular VSD Occluder is intended in patients for transcatheter closure of a complex ventricular septal defect of a sufficient size to warrant closure (large volume left to right shunt, pulmonary hypertension and/or clinical symptoms of congestive heart failure). The patient must also be considered at high risk for surgical closure based either on the anatomy of the defect or the patient's overall medical condition. VSDs typically occur in infants and are the most common type of congenital heart defect.

"We are pleased to make the Muscular VSD Occluder available for our U.S. physician customers," said Franck Gougeon, President and CEO of AGA. "The VSD, along with the previously approved AMPLATZER Septal Occluder and

AMPLATZER Duct Occluder, provide U.S. cardiologists with the most comprehensive set of tools from any single company to treat structural heart disease."

The AMPLATZER Muscular VSD Occluder consists of two discs made of self expanding nitinol. The device is implanted via a catheter, a procedure that is a less-invasive alternative to open heart surgery and results in shorter recovery time. To increase the closing ability, the discs are filled with polyester fabric that is secured to the disc. The Muscular VSD Occluder uses the unique AMPLATZER interface between the device and delivery cable. This screw attachment permits the device to be retrieved and repositioned prior to release from the cable.

As part of the approval conditions, AGA has agreed to conduct a five-year post approval study to evaluate long-term safety and effectiveness. In addition, AGA will enroll at least 100 patients in a prospective registry study that will include an evaluation of immediate post-procedural technical success, occurrence of major complications acutely and over designated time intervals, as well as longer term success in closing the shunt associated with the VSD.

AGA Medical Corporation, based in Plymouth, Minnesota (just outside Minneapolis) is the leader in developing interventional devices to treat structural heart defects. As a result of the many contributions and creative genius of Dr. Kurt Amplatz, the company has developed and commercializes a series of devices that have revolutionized the treatment of the most common congenital "holes in the heart" such as atrial septal and patent foramen ovale defects. The company is expanding into new areas such as the minimally invasive repair of vascular abnormalities. AGA Medical devices have received regulatory approval and are marketed in over 90 countries with over 250,000 devices shipped to date. For more information visit www.amplatzer.com.

Drug Used for Treatment for Heart Failure in Adults May Not be Beneficial for Children and Teens

Preliminary findings indicate a heart failure medication used by adults, carvedilol, may not significantly improve heart failure outcomes for children and adolescents, according to an article in the September 12 issue of JAMA.

"Heart failure due to systemic ventricular dysfunction is a significant medical problem for children and represents the reason for at least 50% of pediatric referrals for heart transplantation. To date, there have been no large randomized controlled trials of any medication in children and adolescents with chronic heart failure. Treatment recommendations in children and adolescents with heart failure are extrapolated from the results of clinical trials conducted in adults, which may be problematic," the authors write.

Robert E. Shaddy, M.D., of Children's Hospital of Philadelphia and the University of Pennsylvania, and colleagues evaluated the effects of the beta-blocker carvedilol in 161 children and adolescents with heart failure. In addition to treatment with conventional heart failure medications, patients were randomized to receive placebo or carvedilol for eight months. The size of the dosage was determined by the weight of the child.

The researchers found no statistically significant difference between the treatment groups with regard to the percentage of patients who improved, worsened, or were unchanged during the course of the study. Among 54 patients assigned to placebo, 56% improved, 30% worsened and 15% were unchanged. Among 103 patients assigned to carvedilol, 56 improved, 24% worsened and 19% were unchanged.



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"This study did not detect a treatment effect of carvedilol on the primary composite end point of clinical heart failure outcomes. It is possible that children and adolescents with heart failure do not receive benefit from carvedilol; this would represent the first heart failure population not to show benefit with beta-blockade and is inconsistent with the many small studies supporting the benefit of beta-blockade in this patient population to date. It is unclear why carvedilol would be beneficial in adults with heart failure but not in children and adolescents," the authors write. "... given the lower than expected event rates, the trial may have been underpowered. There may be a differential effect of carvedilol in children and adolescents based on ventricular morphology."

For more information www.jamamedia.org.

New Digital Grid Will Link Heart Researchers Worldwide

Federally Funded Project Will Allow International Access to Cardiovascular Medical Data Supported by an \$8.5 million federal grant, leading researchers at three universities, including Johns Hopkins, are creating an ambitious digital network that will allow cardiovascular researchers worldwide to easily exchange data and expertise on heart-related illnesses. The project, called the Cardiovascular Research Grid, is expected to be a boon to the large community of heart researchers who will use these digital tools to find new ways to prevent, detect and treat life-threatening cardiac ailments.

To launch this effort, the National Heart, Lung and Blood Institute, part of the National Institutes of Health, has approved an \$8.5 million grant to be allocated over a four-year period that began March 1. The digital project will be based at the Institute for Computational Medicine at Johns Hopkins, in collaboration with the Department of Biomedical Informatics at Ohio State University College of Medicine and the Center for Research in Biological Systems, University of California, San Diego.

The project teams will develop open, grid-based software tools that will enable other research groups to become a "node" in the new grid. Once connected to the grid, researchers will be able to access and share experimental data, data analysis tools and computational models relating to heart



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Surgical procedures include the repair or palliation of all forms of congenital heart defects, including tetralogy of fallot, transposition of the great arteries, hypoplastic left heart syndrome (HLHS) and other forms of single ventricle disease. Furthermore, cardiac transplantation will be available for children with end-stage or non-correctable heart disease. In addition to these complex techniques, we perform minimally invasive endoscopic surgeries with an eye to the future of using robotic surgeries. By virtue of fetal echocardiography and our state-of-the-art imaging center, we also offer a full array of cutting-edge, non-invasive diagnostic techniques, including Cardiac MRI and 3D echocardiography.



Medical City Hospital has been in the heart of Dallas since 1974. Medical City Children's Hospital offers every pediatric specialty and subspecialty and treats everything from the routine to the unexpected. Many of the brightest minds in medicine walk our brightly colored halls daily, enjoying the benefits of working in an innovative, nurturing atmosphere. Major capital expansion of the current hospital facilities includes a new Children's Hospital tower to open in 2008. With desirable market demographics, an excellent payor mix, and a growing market, it's easy to see why physicians come to practice here.

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function in health people and those with cardiac disease. To protect privacy, none of the heart data will carry information identifying patients from whom it was obtained. "There had never been a simple and direct way for cardiovascular researchers to share, analyze and model this important data," said Raimond Winslow, Director of the Institute for Computational Medicine at Johns Hopkins and principal investigator in the project. "Now, there will be."

Winslow, who also is a professor in the Department of Biomedical Engineering, added, "This is the direction in which biomedical research is heading in the 21st Century. In the past, biomedical research was mainly done in individual labs. The Cardiovascular Research Grid will enable us to assemble large, geographically distributed research teams and bring together the leading experts in the world to focus on a common problem, regardless of their location. This grid will enable experimentalists to share their data with computational scientists, who will analyze and model the data. The computational scientists will then share their results with their experimental colleagues who use it to refine their experiments. In this fashion, we believe the creation of the Cardiovascular Research Grid will accelerate the discovery of new approaches for treating heart disease."

In deciding to fund the new grid, the National Heart, Lung and Blood Institute recognized the important contribution that bioinformatics can now make in developing a deeper understanding of the mechanisms of heart disease and in the development of new therapeutic approaches. During the first year of funding, the organizers of the new grid plan will deploy the initial infrastructure and software that will enable researchers to begin sharing and analyzing information. To accomplish this, Joel Saltz, Chair of the Department of Biomedical Informatics and the Davis Endowed Chair of Cancer at Ohio State University, and his team will develop the software infrastructure that ties together resources on the grid. "The Cardiovascular Research Grid will allow experts from different disciplines to combine their insights and to coordinate their efforts," Saltz said. "The ability to bring together many types of biomedical information will have a tremendous impact on the pace of progress in cardiovascular research."

The Johns Hopkins team will focus on development of standardized vocabularies for describing biomedical data, models and data analysis applications. In addition to Winslow, the team will include faculty members Michael I. Miller and Tilak Ratnanather from the Department of Biomedical Engineering; and Donald Geman, Daniel Naiman and Laurent Younes, all from the Department of Applied Mathematics and Statistics. Mark Ellisman, Director of the National Center for Microscopy and Imaging at the University of California, San Diego, and his team will be responsible for developing effective and intuitive ways for users to interact with the Cardiovascular Research Grid.

"Developing and deploying cyberinfrastructure to capitalize on emerging technologies to promote better collaboration and accelerate research is a core focus of our Center's efforts," said Ellisman, who also is Director of UCSD's Center for Research in Biological Systems. "With a track record of developing scalable cyberinfrastructure to foster interdisciplinary investigations among teams of researchers in microscopy, neuroimaging and the environmental health sciences, CRBS is eager to collaborate with the Johns Hopkins team on developing the Cardiovascular Research Grid. We're looking forward to implementing an infrastructure that will effectively pool the diverse expertise, applications and instrumentation of the cardiovascular research community into a unified knowledge base—one that will enable researchers to tackle cardiac disease studies of greater scope and complexity."

The Cardiovascular Research Grid will be headquartered in the 79,000 square-foot Computational Science and Engineering Building, now under construction on the Homewood campus of Johns Hopkins. The building is expected to open this summer.

For more information, www.jhu.edu/news_info/news.



Director of Pediatric Cardiovascular Critical Care

Major Medical Center in Dallas seeks a Pediatric Cardiovascular Critical Care Director. Preferred candidate will possess a charismatic personality, leadership attributes with evidenced experience, strong clinical skill set for a complex patient population and board certification in Pediatric Cardiology and Critical Care. Candidates with board certification in one discipline and solid experience in the alternate subspecialty should also apply. The incoming Director will serve as the Medical Director of the existing 10 bed Pediatric Cardiovascular ICU and the new, state-of-the-art unit due for completion in late 2008. Additional responsibility includes coordinating a collegial collaboration with pediatric cardiology physicians/subspecialists and nursing staff. Incoming physician will be provided an outstanding financial package and the opportunity to advance their medical and/or research career.

The Congenital Heart Surgery program performs more than 300 surgeries each year. Two thirds of the surgeries are pump cases. The program provides care to neonates (approximately 30%) and children under 2 yrs of age (approximately 70%).

A team of nine pediatric intensivists and eleven pediatric cardiologists cover the congenital heart surgery unit. A dedicated 10-bed pediatric cardiovascular intensive care unit opened in 2004. Construction has begun on a newer unit with a completion date in late 2008.

The program participates extensively in research initiatives and i-Rounds, a web based informatics system allowing second to second tracking of clinical data and shares information with outside referring physicians. The center employs all of the latest technologies for monitoring patients and performing point of care testing. The Medical Center operates a very busy research entity in which the Pediatric Cardiovascular Surgery Director, actively participates.

Call or inquire by email today:
Kathleen Kyer, Manager,
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888-933-1433 or
Kathleen.Kyer@HCAHealthcare.com

PICS 2007 Wrap-up

By Ziyad M. Hijazi, MD

The Pediatric Interventional Cardiac Symposium (PICS) was held in Las Vegas at the Bellagio Hotel from July 22-25, 2007. This year over 800 professionals from over 50 countries attended the three-and-a-half day symposium. This year, the meeting had over 110 distinguished faculty from around the world.

The meeting was dedicated to the memory of Mr. Joel Sims, a cardiovascular technologist who worked at the University of Chicago for 20 years. Joel was intimately involved with the organization of PICS since Dr. Hijazi moved to Chicago in 1999, and he had worked on all meetings until his death earlier this year. He will be missed very dearly.

The 2007 meeting started Sunday, July 22nd, in the afternoon, where the first session was a workshop on ASD/PFO closure and imaging of the atrial septum. Various devices and imaging modalities were discussed, including the Helex device, the Occlutech device, the AcuNav, and the new 3D TEE imaging for the septum. The workshop was followed by oral abstract presentations in three different simultaneous rooms. Thirty abstracts were presented in these rooms. The abstracts were the best of over 100 submitted to the meeting. The last session was "Meet the Expert" where attendees brought difficult and challenging cases which were discussed with the course distinguished faculty. At the end of the day, a welcome reception took place at the opening of the exhibit. Over 30 exhibitors representing various device/catheter/publishing/organization (Society) had booths at the meeting.

Monday July 23rd, 2007 was the official day of opening of the meeting. The course's director delivered his annual speech and went over the program. Then

attendees enjoyed a very busy day full of lectures and live cases in two different arenas. The Pediatric arena focused on con-



Dr. Terry King being presented with the first "Pioneer Award." Left to right: Drs. William E. Hellenbrand, Ziyad M. Hijazi and Terry D. King.

genital heart disease in children and adults and the adult arena focused mainly on structural heart disease as well as on congenital heart disease in the adult patients. Live cases were transmitted from Milan by Drs. Mario Carminati and Gianfranco Butera, Sao Paulo by Drs. Carlos Pedra and Cesar Esteves, Toronto Sick Kids by Dr. Lee Benson, New York City by Dr. Robert Sommer and Seattle by Drs. Mark Reisman and Tom Jones. Some new devices were featured for the first time including the Cardia Intrasept device for ASD closure and the Occlutech device for PFO closure. The quality of the cases was excellent and the attendees enjoyed the direct interactions with the operators. For the first time this year, Dr. Lee Benson gave a live tour of his magnificent catheterization suites!

On Monday, two awards were given. The first one was "The Pioneer Award" which

was given to Dr. Terry D. King who was the first in the world to close an ASD percutaneously in April 1975. Dr. Hijazi went over the life and accomplishments of Dr. King including showing of the news clip from NBC when he performed the first case. Dr. King's family surprised him and flew to attend the ceremony without him knowing about it. The second award was the usual "PICS Achievement Award." This year's recipient of the award was Dr. John P. Cheatham for his innovations and contributions to the ever-changing field of interventional pediatric cardiology. Dr. Hijazi went over Dr. Cheatham's life and showed photos of John in his various stages of his career.

Tuesday, July 24th, 2007 again featured many live cases transmitted from Sunrise hospital in Vegas where Drs. Al Galindo, Abe Rothman and Ziyad M. Hijazi performed three very good cases, St. John's Hospital in Phoenix where Drs. Steve Pophal and John P. Cheatham performed three cases, Omaha Children's Hospital where Dr. Zahid Amin performed 2 cases, and St. Paul Hospital where Dr. John



Dr. John P. Cheatham being presented with the "PICS Achievement Award." Left to right: Drs. John P. Cheatham, Ziyad M. Hijazi and William E. Hellenbrand.



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“Preparations are well underway for next year’s meeting (PICS-XII) also to be held at the Bellagio in Las Vegas, July 20th-23rd, 2008. Already, the following institutions have confirmed their participation as live case sites....”

Webb performed two cases, one of them was a percutaneous aortic valve implantation case. In addition to the live cases, didactic lectures were given in both rooms discussing various topics in congenital and valvular/structural heart disease.

The last day featured a combined session between adult and pediatric cardiologists where the emphasis was on adult congenital heart disease including a session on the right ventricle outflow tract. Live cases that day were transmitted from Miami Children’s Hospital where Drs. Evan Zahn, Redmond Burke and Bob Hannan performed excellent hybrid cases. For the first time the transmission from Miami was HD, and the attendees were very impressed by the quality of images. Also, Dr. David Balzer from St. Louis Children’s Hospital performed three cases and Dr. Eric Horlick with Dr. Lee Benson performed cases from Toronto General Hospital. Also for the first time, Drs. Horlick and Benson performed a live case closing a PFO using the new bioabsorbable device “BioSTAR” from NMT.

In addition to the main sessions in both rooms, breakout sessions for technologists and nurses took place simultaneously to discuss issues related to their field.

At the end of the meeting on Wednesday, the Gala dinner took place at the Bellagio where attendees relaxed and enjoyed the evening.

Preparations are well underway for next year’s meeting (PICS-XII) also to be held at the Bellagio in Las Vegas, July 20th-23rd, 2008. The following institutions have already confirmed their participation



Dr. Hala Al Kherbash, winner of Congenital Cardiology Today’s drawing for an Apple iPod.

as live case sites: Rush University Medical Center (Operators: Ziyad M. Hijazi and Zahid Amin); Columbus Children’s Hospital (Operators: John P. Cheatham, Mark Galantowicz, Ralf Holzer and Sharon Hill); Danta Pazzanese in Brazil (Operators: Carlos Pedra and Cesar Esteves); Miami Children’s Hospital (Operators: Evan Zahn and Redmond Burke); St. Louis Children’s Hospital (Operator: David Balzer); Seattle Children’s Hospital (Operator: Tom Jones); Detroit Children’s Hospital (Operator: Tom Forbes); Orlando Regional Children’s Hospital (Operator: David Nykanen); Emory Children’s Hospital (operator: Robert Vincent); Cincinnati Children’s Hospital (Operators: Russell Hirsch & Robert Beekman), and perhaps two other sites are being considered. We promise you that it will be an excellent educational opportunity for you and your staff. We hope to see you there.

For more details on PICS 2008, please visit the PICS website at: www.picsymposium.com.

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On Behalf of PICS Course Directors & Co-Directors

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CONGENITAL CARDIOLOGY TODAY

© 2007 by Congenital Cardiology Today
ISSN: 1544-7787 (print); 1544-0499 (online).
© 2007 by Congenital Cardiology Today
(ISSN 1544-7787-print; ISSN 1544-0499-online).
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