

# CONGENITAL CARDIOLOGY TODAY

RELIABLE INFORMATION IN CONGENITAL CARDIOLOGY

INTERNATIONAL EDITION

WWW.CONGENITALCARDIOLOGYTODAY.COM

VOL. 3, ISSUE 7 JULY 2005

Formerly Pediatric Cardiology Today

## INSIDE THIS ISSUE

**Patent Ductus Arteriosus Stenting – Problems, Complications and Technical Consideration** 1  
*by Mazeni Alwi, MD*

**Tackling the Problem of "Atypical" Kawasaki Disease: A Commentary on the Recently Published AHA Statement for Health Professionals** 6  
*by Masato Takahashi, MD*

**Highlights from the 28th Annual Scientific Session of the Society for Cardiovascular Angiography and Interventions** 11  
*by José A. Ettedgui, MD*

**Paediatric Cardiology Loses A Pioneer: Robert M. Freedom MD, FRCP(C), FACC, O Ont 1941-2005** 15  
*by Joel A. Kirsh, MD; John Pires; Lee N. Benson, MD*

## DEPARTMENTS

**Medical Conferences** 5

**Useful Medical Websites** 10

**Medical News and Information** 14

## CONGENITAL CARDIOLOGY TODAY

9008 Copenhagen Drive, Ste. M  
Potomac, MD 20854 USA  
[www.CongenitalCardiologyToday.com](http://www.CongenitalCardiologyToday.com)

© 2005 by Congenital Cardiology Today (ISSN 1554-7787-print; ISSN 1554-049 - online). Published monthly. All rights reserved. Statements or opinions expressed in Congenital Cardiology Today reflect the views of the authors and are not necessarily the views of Congenital Cardiology Today.

## PATENT DUCTUS ARTERIOSUS STENTING – PROBLEMS, COMPLICATIONS AND TECHNICAL CONSIDERATION

*By Mazeni Alwi, MD*

Advances in surgical techniques and post operative care have significantly changed the management of cyanotic congenital heart disease. The trend is towards definitive repair in early infancy as far as possible, obviating the need for palliative surgery as first stage treatment in many conditions. This is best illustrated by the way non duct-dependent Tetralogy of Fallot (TOF) is managed in the modern era, where total correction is often performed during mid to late infancy, such that palliative modified Blalock -Taussig shunt (BTS) is rarely performed except in the very small symptomatic patients. However, the BTS, as a bridge towards definitive repair still forms an important part in the management of the more complex cyanotic heart disease, particularly for those in whom the pulmonary blood flow is duct-dependent. This applies to diverse lesions, from TOF to hearts with single ventricle physiology where pulmonary atresia is the unifying thread. Because these are duct-dependent lesions, BTS in the modern era is almost exclusively performed in the neonatal period. Compared to BTS performed later in childhood, those performed in the neonatal period are not unexpectedly associated with high morbidity. The early complications include early shunt failure due to acute thrombosis, overshunting with heart failure features, chylothorax, pleural effusions and diaphragmatic paralysis. There is especially increased mortality in patients with pulmonary atresia with intact ventricular septum (PA-IVS) with the added feature of major coronary sinusoids. Because the shunted branch pulmonary arteries are often small, no bigger than 4 mm in most cases, the late complications of pulmonary artery distortion or stenosis is of concern, especially those destined for the Fontan track.

Because of these issues related to neonatal

shunts, there have been a number of novel attempts to maintain ductal patency for a longer term as an alternative form of palliation in these duct-dependent lesions. Formalin infiltration of the patent ductus arteriosus (PDA) at thoracotomy was one of the earliest suggestions but this is almost just as invasive as conventional shunt. Oral Prostaglandin has also been advocated but the frequency at which it has to be administered makes this impractical whilst there was also concern about its efficacy. With advances in transcatheter balloon dilatation techniques in the neonates, simple balloons or balloon heated with laser or radiofrequency has been suggested, but again the problem of efficacy or consistency of results arose<sup>1</sup>.

The real prospect of transcatheter manipulation of the ductus arteriosus to reliably maintain its patency became possible when balloon expanded coronary stents came into wide clinical use as an extension of PTCA in the management of CAD. In an experimental study involving newborn lambs, Coe et al successfully implanted stainless steel stents without encountering major complications, achieving continued ductal patency for up to 3 months when

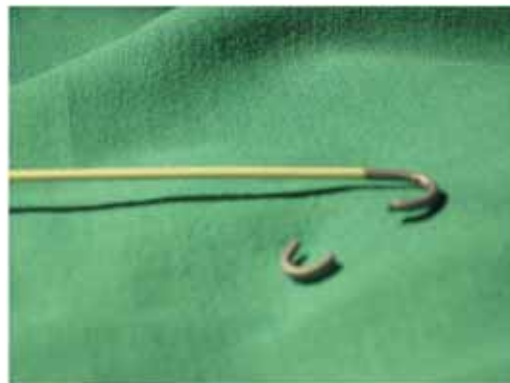


Figure 1. Cut pigtail for engagement of ducts with proximal origin

# CONGENITAL CARDIOLOGY TODAY

Formerly Pediatric Cardiology Today

© 2005 by Congenital Cardiology Today  
(ISSN 1554-7787 - print; ISSN 1554-049 - online)  
Published monthly. All rights reserved.

## Publishing Management

Tony Carlson, Founder  
TonyC@CCT.bz

Richard Koulbanis, Publisher & Editor  
RichardK@CCT.bz

John W. Moore, MD, MPH, Medical Editor & Medical Editorial Board  
jwmoore@mednet.ucla.edu

## Editorial Board

Zohair Al Halees, MD

Mazeni Alwi, MD

Felix Berger, MD

Fadi Bitar, MD

Philipp Bonhoeffer, MD

Anthony C. Chang, MD, MBA

Bharat Dalvi, MD, MBBS, DM

Horacio Faella, MD

Yun-Ching Fu, MD

Felipe Heusser, MD

Ziyad M. Hijazi, MD, MPH

Ralf Holzer, MD

R. Krishna Kumar, MD, DM, MBBS

Gerald Ross Marx, MD

Tarek S. Momenah, MBBS, DCH

Toshio Nakanishi, MD, PhD

Carlos A. C. Pedra, MD

James C. Perry, MD

Shakeel A. Qureshi, MD

Andrew Redington, MD

Carlos E. Ruiz, MD, PhD

Girish S. Shirali, MD

Horst Sievert, MD

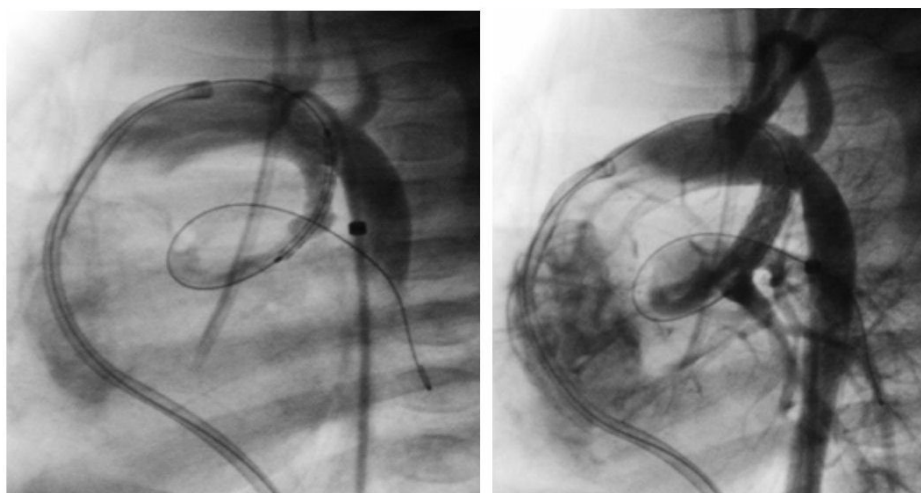
Hideshi Tomita, MD

Gil Wernovsky, MD

Carlos Zabal, MD

## To Contact an Editorial Board Member

Email to: NAME@CCT.bz. Place the Board Member's surname before the @.



a

b

Figure 2. Tetralogy of Fallot with pulmonary atresia, proximal origin of PDA. Stent implantation by transvenous route through the VSD. Balloon and stent across the PDA (a). Stent fully expanded (b).

they were sacrificed<sup>2</sup>. However, transferring PDA stenting from the animal laboratory to clinical practice has met with several major problems.

The early report of PDA stenting in duct dependent cyanotic heart disease by Gibbs highlighted these difficulties and potential complications of this procedure in neonates. Obviously small patient size and, therefore, small vessels, cardiac chambers and thin walls are susceptible to damage and perforation with manipulation of stiff catheters and guide wires. Moreover, the stent and balloon during that era required a 5F or 6F sheath for delivery, making the transfemoral arterial route not possible.<sup>3</sup> But more than just patient size, the PDA in cyanotic duct-dependent lesion are often longer, more tortuous and arise more proximally from the aorta. This is especially so in TOF with pulmonary atresia (PA) where the PDA may arise very proximally from the underside of the arch, giving the appearance of a vertical duct. This poses a major technical problem when stenting is

attempted via the femoral arterial route due to the difficulty of engaging the ductal ampulla and getting a stable wire position for tracking of balloon and stent. For these two reasons, PDA stenting in these duct-dependent neonates were performed via axillary artery cut-down in the earlier reports.

The additional difficulties that were encountered in this early experience were the inability to enter the duct, ductal spasm and incomplete stenting of the full length of the duct, hence requiring repeat procedures. It is hardly surprising that after 6 years with 7 successful procedures out of 11 patients with duct-dependent pulmonary circulation (most probably not all consecutive patients were recruited) with 2 failures and 2 deaths due to ductal spasm, apart from poor duration of palliation due to endothelial proliferation, the authors concluded that ductal stenting in this group of patients cannot be recommended<sup>4</sup>.

However, Schneider et al reported a more encouraging outcome of PDA



## PICS/ENTICHS- 2005

Pediatric Interventional Cardiac Symposium

and Emerging New Technologies in Congenital Heart Surgery

SEPTEMBER 15-18, 2005 At the Hilton Buenos Aires Buenos Aires, Argentina

Immediately preceding the 4th World Congress of Pediatric Cardiology and Cardiac Surgery

[www.picsymposium.com](http://www.picsymposium.com)

stenting in neonates with duct dependent pulmonary circulation performed within the similar period<sup>5</sup>. The authors divided the patients into 2 groups, the first (8/21) were those with critical pulmonary stenosis (PS) or pulmonary atresia intact ventricular septum (PAIVS) who primarily underwent transcatheter relief of right ventricular outflow obstruction but later required additional source of pulmonary blood flow due to mainly small right ventricular size/poor right ventricular compliance. In this group, PDA stenting was performed transvenously via the newly opened pulmonary valve. The other group (13/21) was comprised of patients with pulmonary atresia in the setting of TOF, tricuspid atresia and transposition of great artery (TGA), ventricular septal defect (VSD). In this second group PDA stenting was performed by the transarterial route. The access was via axillary artery cut-down in the majority of cases.

Similarly, Gewillig et al also reported encouraging results of PDA stenting in patients who required additional source of pulmonary blood flow after relief of right ventricular outflow tract obstruction in critical PS or PAIVS.<sup>6</sup> Clearly, in this group of patients, PDA stenting by the transvenous route is a fairly straightforward procedure once the pulmonary valve is opened.

The current generation of coronary stents, delivery catheters and wires have better flexibility and lower profile that some of the earlier problems of PDA stenting may be overcome. Newer features such as heparin coating and drug elution may be helpful in extending the duration of palliation. It is perhaps timely to examine the problems and technical difficulties and suggest technical improvement of PDA stenting in the other group – pulmonary atresia in the setting of TOF, tricuspid atresia, TGA, VSD and univentricular hearts where access to the

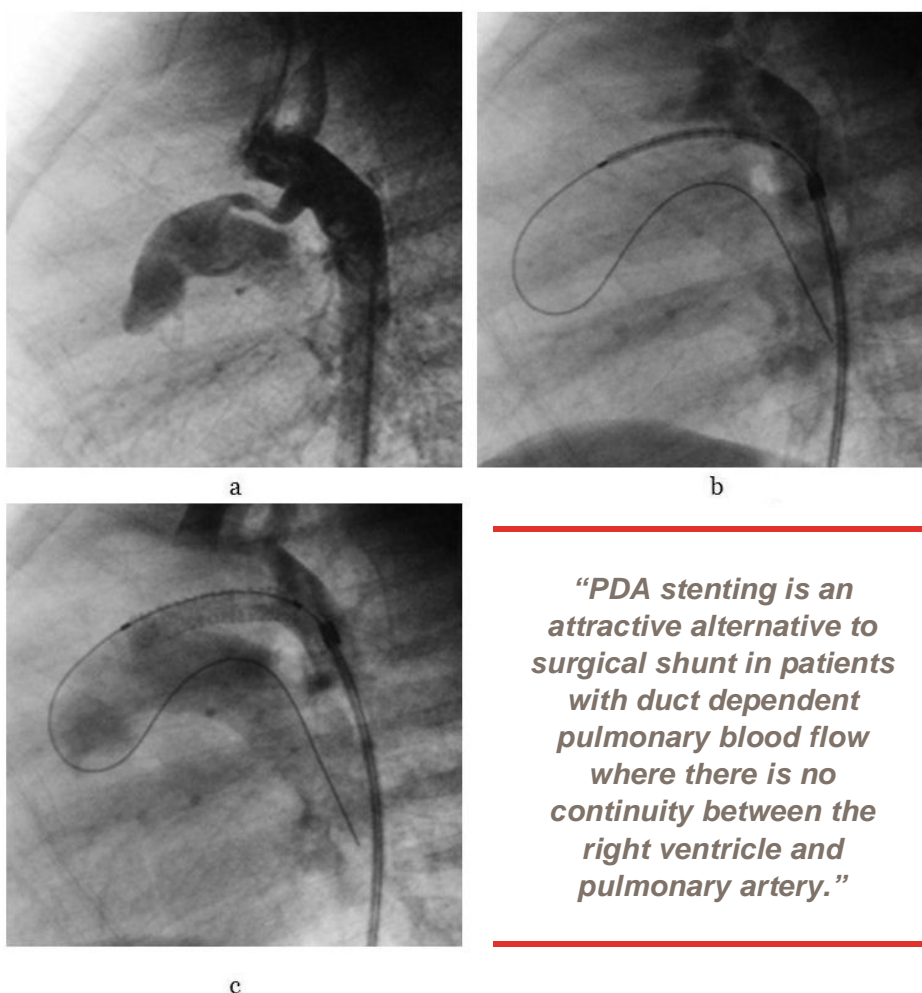


Figure 3. PDA stenting by transfemoral arterial route. Curved PDA (a). Straightens with guide wire (b). Post stent expansion (c).

PDA is only via the aorta given the limitations to vascular access via the femoral arterial route due to small vessel size and the peculiar morphology of the ductus which in these lesions tend to be long, tortuous and arise proximally. In the past, this has been achieved by axillary artery cut-down to engage the ductal ampulla and secure good wire position for stent delivery and deployment. However, most cardiologists today find axillary artery cut-down too invasive and

perhaps a major deterrent to attempts at PDA stenting. Can this be performed via the femoral artery?

The small vessel size is largely overcome with lower profile designs of coronary stents and delivery catheters today which can be delivered via 4F sheath. For this, 4F long sheaths is available by special order (Cook Inc.). The other problem with the transfemoral arterial route is the difficulty of engaging and

---

***“PDA stenting is an attractive alternative to surgical shunt in patients with duct dependent pulmonary blood flow where there is no continuity between the right ventricle and pulmonary artery.”***

---

**AMPLATZER®** Vascular Plug



**Simple. Reliable. Repositionable.**

For more information visit: [www.amplatzer.com](http://www.amplatzer.com) or call: 763-513-9227  
Toll free in the US: 888-546-4407





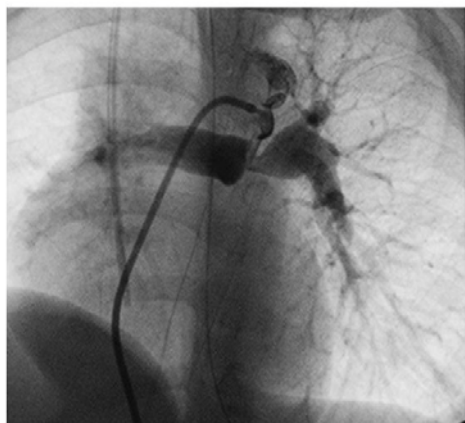


Figure 4. Branch LPA stenosis at the side of ductal insertion in Tetralogy of Fallot with pulmonary atresia.

securing a stable wire position due the proximal origin of the ductus which are often long and tortuous especially in patients with TOF-PA.

This is largely overcome by using a pig-tail catheter with its tip cut off such that it forms the general shape of an "inverted U" (Figure 1). The curve may be refined to suit the origin ("verticalness") of the ductus. In tricuspid atresia and PAIVS, the ductal origin may not be as proximal and the ampulla may thus be engaged easily with a Judkins® catheter. In a small number of cases of TOF-PA, the PDA arises very proximally, such that it is almost impossible to engage the ductal ampulla, allowing only a short length of guide wire to be passed into the pulmonary artery which is certainly insufficient for tracking of balloon and stent. Instead of axillary artery cut-down, the transvenous approach may be used. A 5F Judkins® guiding catheter may be passed from the femoral vein to the right atrium, right ventricle and the aorta via the VSD. (Figure 2) Once the tip is engaged in the ductal ampulla, a stable guide wire position is easier to achieve. The extra-

support choice PT wire (Boston Scientific Inc.) is most useful because it has a short floppy hydrophilic tip which enables crossing of long, tortuous and tight ductus yet has a relatively stiff body, such that once firmly anchored in a branch pulmonary artery, it straightens the ductus arteriosus and provides stability for tracking of stent and balloon catheter (Figure 3).

#### Stent length and size

Extrapolating the experience of Blalock-Taussig shunts, we use mainly 4.0mm, and to a lesser degree, 4.5mm diameter stents. Neo-intimal proliferation and in-stent stenosis develop fairly quickly such that it is wise to plan the second stage operation (Glenn shunt or Rastelli type operation) within 6 – 12 months of PDA stenting, although in a small number of patients good palliation can be achieved beyond 2 years.

Choosing the stent length is more exacting. As a principle, the entire ductus should be stented, otherwise the unstented segment will rapidly constrict and compromise pulmonary blood flow. The exception to this are ducts that arise for the subclavian artery which tend to be very long and have an acute take-off. Length measurement is best taken once the ductus is straightened by having an extra support coronary wire across it, usually using the lateral projection or the 4 chamber view. Stent lengths are chosen to allow 10% shortening with full expansion.

#### Branch pulmonary artery stenosis

The PDA in cyanotic heart disease, apart for its proximal origin, length and tortuosity may be associated with branch pulmonary artery stenosis. In our experience this is especially frequent in TOF- pulmonary atresia, but much less in PAVIS and tricuspid atresia. This is thought to

be due to the extension of ductal tissue into the medium of pulmonary arterial wall, leading to "pulmonary coarctation" when the ductus constricts (Figure 4). We have noted that PDA stenting tends to accelerate pre-existing branch pulmonary artery stenosis, which may require a "salvage" BT shunt on the affected vessel.<sup>7</sup> It is thus important to exclude proximal branch pulmonary stenosis (especially left pulmonary artery) and we consider its presence a contraindication for PDA stenting, especially those destined for the Fontan tract. The 4 chamber view is generally helpful in opening up the pulmonary bifurcation and unmask branch pulmonary artery stenosis at the site of ductal insertion.

#### Complications

Spasm of the patent ductus arteriosus in our experience is surprisingly rare, even in those that are very restrictive. A major, but fortunately rare complication is acute thrombosis where thrombus rapidly fills up a stent within minutes of expansion. This manifests as rapid deterioration in oxygen saturation after an initial excellent improvement. In such a situation, thrombolytic therapy is recommended and should be administered to prepare the patient for BT shunt. Inadvertent dislodgement of stent can be minimized by ensuring stable wire anchoring before stent deployment. Transvenous stenting of TOF-pulmonary atresia with proximal origin of the PDA may cause complete heart block as the stiff coronary guiding catheter presses on the rim of the VSD where the AV node runs.

#### Conclusion

PDA stenting is an attractive alternative to surgical shunt in patients with duct-dependent pulmonary blood flow where there is no continuity between the right ventricle and pulmonary artery. Small

## Do You Want to Recruit a Pediatric Cardiologist?

Advertise in the only monthly publication totally dedicated to pediatric and congenital cardiology. For more information:

[Recruit@CCT.bz](mailto:Recruit@CCT.bz)

femoral artery and the peculiar morphology of the PDA in these lesions present unique difficulties to percutaneous PDA stenting. With the present generation of low profile and more flexible stent and delivery systems, this procedure can be accomplished by the transfemoral arterial route with techniques described above. Occasionally, the transvenous route via the VSD may be required in TOF – PA with very proximal origin of the PDA. Special attention needs to be paid to pre-existing branch PA stenosis at the ductal insertion site which we consider as contraindication to PDA stenting, especially in patients destined for the Fontan track.

# References

1. John L Gibbs. *Stenting the arterial duct*. Archives of disease in childhood, 1995; 72: 196 - 197.
2. James Y. Coe, Peter M. Olley. *A novel method to maintain ductus arteriosus patency*. J Am Coll Cardiol 1991; 18: 837 - 841.
3. John L Gibbs, Martin T Rothman, Michael R Rees, Jonathan M Parsons, Mike E Blackburn, Carlos E Ruiz. *Stenting of the arterial duct: a new approach to palliation of pulmonary atresia*. Br Heart J, 1992; 67: 240 - 245.
4. John L Gibbs, Orhan Uzun, Michael EC Blackburn, Christopher Wren, JR Leslie Hamilton, Kevin G Watterson. *Fate of the stented arterial duct*. Circulation, 1999; 99: 2621 - 2625.
5. M. Schneider, P. Zartner, A. Sidiropoulos, W. Konertz, G. Hausdorf. *Stent implantation of the arterial duct in newborns with duct-dependent circulation*. Eur Heart J 1998; 19: 1401 - 1409.
6. Marc Gewillig, Derize E Boshoff, Joseph Dens, Luc Mertens, Lee N Benson. *Stenting the neonatal arterial duct in duct-dependent pulmonary circulation: new*

*techniques, better results*. J Am Coll Cardiol 2004; 43: 107 - 112.

7. Mazeni Alwi, KK Choo, Haifa Abdul Latiff, Geetha Kandavello, Hasri Samion, MD Mulyadi. *Initial results and medium-term follow up of stent implantation of patent ductus arteriosus in duct-dependent pulmonary circulation*. J Am Coll Cardiol 2004; 44: 438 - 445.

Figures 2 and 4. Mazeni Alwi, KK Choo, Haifa Abdul Latiff, Geetha Kandavello, Hasri Samion, MD Mulyadi. *Initial results and medium-term follow up of stent implantation of patent ductus arteriosus in duct-dependent pulmonary circulation*. J Am Coll Cardiol 2004; 44: 438 - 445. (c) 2004 Reprinted with permission from The American College of Cardiology Foundation.

Email comments on this article to:  
JULYMA@CCT.bz

~CCT~



Mazeni Alwi, MD  
Consultant & Head  
Department of Paediatric Cardiology  
Institut Jantung Negara (National Heart Institute)  
Kuala Lumpur  
muhaneeza@ijn.com.my

**Correction to the June issue of Congenital Cardiology Today (CCT), page 12, the September Conference Focus - PICS/ENTICHS 2005.** CCT should have listed the Course Directors for PICS/ENTICHS 2005 as Ziyad M. Hijazi, MD, MPH and William E. Hellenbrand, MD. For more information see [www.picsymposium.com](http://www.picsymposium.com)

## MEDICAL CONFERENCES

**12th World Congress on Heart Disease**  
July 16-19, 2005; Vancouver, Canada  
[www.cardiologyonline.com](http://www.cardiologyonline.com)

**SIS (Summer in Seattle) 2005**  
July 19-23, 2005; Seattle WA USA  
[www.summerinseattle.com](http://www.summerinseattle.com)

**New Concepts in Exercise and Pulmonary Hypertension in the Young: Their Interaction at Sea Level and Altitude**  
August 5-7, 2005; Aspen CO USA  
Taylor.Amy@tchden.org or  
[www.congenitalcardiology.com/Cardiac-Aspen2005.pdf](http://www.congenitalcardiology.com/Cardiac-Aspen2005.pdf)

**IPTA's (International Pediatric Transplant Association) 3rd World Congress on Pediatric Transplantation Medicine!**  
August 6-9, 2005; Innsbruck, Austria  
[www.IPTAonline.org](http://www.IPTAonline.org)

**British Society Cardiovascular Research - "Stress Signals in the Cardiovascular System"**  
Sep. 15-16, 2005; London, UK  
[www.bscr.org](http://www.bscr.org)

**PICS-IX and ENTICHS-III (9th Pediatric Interventional Cardiac Symposium & Third Emerging New Technologies in Congenital Heart Surgery)**  
Sep. 15-18, 2005; Buenos Aires, Argentina  
[www.picsymposium.com](http://www.picsymposium.com)

**The 4th World Congress of Pediatric Cardiology and Cardiac Surgery**  
Sep. 18-22, 2005; Buenos Aires, Argentina  
[www.pccs.com.ar/](http://www.pccs.com.ar/)

**9th Annual Meeting of the Midwest Pediatric Cardiology Society**  
Oct. 20-21, 2005; Iowa City IA USA  
[www.mwpcsociety.org](http://www.mwpcsociety.org)



[www.numed.on.ca](http://www.numed.on.ca)

**NuMED, Inc.**

Hallenweg 40, 5683 CT Best, The Netherlands  
Tel: +31 (0)499 377388 Fax: +31 (0)499 377456

Manufacturer of angioplasty and valvuloplasty catheters, has a long standing commitment in meeting our customer's expectations by providing a high quality product. At NuMED, we see quality improvement as a continual process, aimed at satisfying these expectations and requirements at every stage.

## TACKLING THE PROBLEM OF "ATYPICAL" KAWASAKI DISEASE: A COMMENTARY ON THE RECENTLY PUBLISHED AHA STATEMENT FOR HEALTH PROFESSIONALS

By Masato Takahashi, MD

**Background:** It has been 38 years since the original description of Kawasaki Disease (KD). KD is no longer a rare or uncommon disease. An average pediatric practitioner encounters this entity repeatedly in his/her practice. It is the leading cause of acquired heart disease in children. Current incidence of KD in Los Angeles County is 23 per 100,000 children under 5 years of age. Although advances have been made in understanding the epidemiology, pathogenesis and acute phase treatment of this disease, we still do not know the etiologic agent(s). Lacking in a fail-safe diagnostic test, we continue to rely on the clinical criteria, which are almost identical to the ones originally proposed by Dr. Kawasaki. Diagnosis is often missed or delayed, if the patient is outside the usual age bracket (6 months to 5 years); if the symptoms appear one by one over a span of 7 to 10 days rather than within one or two days; or if the symptoms are too few in numbers to fulfill the standard diagnostic criteria. If diagnosis is delayed beyond the 10th illness day, there is an increased probability of coronary artery aneurysm formation.

For most cases of incomplete (atypical) KD, usually a physician in a tertiary care facility ends up "breaking the news" to the family about the diagnosis. Having spent sleepless nights with the sick child with a diagnosis of "viral syndrome," having made several futile outpatients visits, and having tried a

succession of ineffective courses of antibiotics, the parents are now con-

fronted with a dreaded coronary artery disease. Their frustration can easily

Evaluation of Suspected Incomplete Kawasaki Disease (KD)<sup>1</sup>



1 This algorithm is not evidence-based, but represents consensus of experts.

2 Exception: infants ≤ 6 months old with ≥ 7 days of fever without plausible cause should undergo laboratory testing. If there is evidence of systemic inflammation, echo should be done even if the infant has no clinical criteria.

Figure 1. Evaluation of suspected incomplete Kawasaki Disease. In the absence of a gold standard for diagnosis, this algorithm cannot be evidence-based but rather represents the informed opinion of the expert committee. From *Circulation* with permission



### The Fourth World Congress of Pediatric Cardiology and Cardiac Surgery

Buenos Aires, Argentina • September 18-22, 2005

[www.pccs.com.ar](http://www.pccs.com.ar)



turn into anger toward the care providers who “missed” the diagnosis, which seems obvious in hindsight. Such a scenario often leads to malpractice litigation.

Although the classic diagnostic guidelines (Table 1)<sup>1</sup> have stood the test of time as epidemiological case definition, they have not been useful as a tool for early diagnosis and treatment in order to minimize the coronary artery complications, especially for patients with incomplete (atypical) KD, which by definition have already developed coronary artery abnormalities. This is not acceptable in this age of effective IVIG treatment to prevent such abnormalities. Incomplete (atypical) cases currently constitute 15-20% of the total cases.

The revised “statement for health professionals on diagnosis, treatment and long-term management of Kawasaki Disease,” was recently completed by the AHA Committee on Rheumatic Fever, Endocarditis and Kawasaki Disease, and was published in both *Circulation* and *Pediatrics*<sup>2</sup>. The document can also be downloaded from the website: [www.pediatrics.org/cgi/content/full/114/6/1708](http://www.pediatrics.org/cgi/content/full/114/6/1708). This large document combines and updates the topics from previously published guidelines for diagnosis and treatment<sup>1</sup> and those for

long-term management.<sup>3</sup> By co-publishing this document in *Pediatrics*, the writing group sought not only to disseminate the updated information widely, but also to raise the general awareness of incomplete (atypical) Kawasaki Disease among the specialists and primary care physicians alike in order to promote timely diagnosis and treatment and reduce the prevalence of cardiac sequela. The centerpiece of this document is the algorithm (Figure 1) describing a step-by-step evaluation using the readily available clinical tools, including:

- (1) patient's clinical characteristics
- (2) acute phase reactants (ESR and CRP)
- (3) supplementary laboratory tests
- (4) echocardiogram.

When the diagnosis is reasonably certain on clinical and laboratory grounds, the treatment with IVIG may be started without waiting for an echocardiogram. However, when the diagnosis is still in doubt in the light of clinical and laboratory findings, echo may offer an additional diagnostic support, utilizing the expanded array of “positive” findings. The algorithm also incorporates feedback loops for yet undiagnosed patients to capture those cases whose disease progression is slower than usual.

#### Details of the diagnostic algorithm (Figure 1):

**Step 1:** If the patient has > 5 days of fever and only 2 or 3 of the classic clinical criteria, the patient's clinical characteristics are assessed. Here, it is important to recognize not only those clinical characteristics which are consistent with KD, but also those which are inconsistent with KD. The features

in the latter category include:

- *Exudative* conjunctivitis
- *Exudative* pharyngitis
- *Discrete* intraoral lesions
- Bullous / vesicular rash
- Generalized adenopathy (as opposed to a large unilateral cervical node)

A patient with one or more of the above inconsistent characteristics is unlikely to have KD. However, if the fever persists for a few more days, this patient should be reevaluated anew, because he/she may be one with exceptional symptomatology or may have some other unrelated disease.

**Step 2:** If the patient's clinical characteristics are consistent with KD but insufficient in number of criteria, a number of laboratory tests may be of value, starting with acute phase reactants: erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). If CRP is > 3 mg/dL and/or ESR is > 40mm/hr, then a battery of 6 other laboratory results should be examined: serum albumin, hemoglobin and hematocrit, alanine aminotransferase (ALT), platelet count, total WBC and urine microscopic examination for the following criteria of abnormalities:

- Serum albumin < 3 g/dL
- Anemia for age
- Elevated ALT
- Platelets > 450,000 /cu mm after 7 days of illness
- WBC > 15,000 /cu mm
- Urine > 10 WBC / HPF

If 3 or more of these abnormal findings

***“The new AHA statement is a large document incorporating previous two documents on diagnosis and treatment and long-term management.”***



English - Spanish - Italian - Portuguese - French

Intra-departmental • Institutional • Global

[www.PedCath.com](http://www.PedCath.com)

Scientific  
Software  
Solutions

are present, then the diagnosis of KD is very likely and the IVIG plus aspirin treatment should be started. Echocardiogram should be obtained, but is not necessary before starting the treatment.

**Step 3:** If CRP and/or ESR is elevated, but there are not enough of abnormal supplementary laboratory tests, then echo should be done as an additional diagnostic aid. Here, echocardiogram is considered "positive," if one of the following three findings is present (Expanded definition of "positive" echocardiogram for Kawasaki Disease):

- Z value of the internal diameter of the left anterior descending artery (LAD) or the proximal right coronary artery (RCA) is  $> 2.5$ .<sup>4</sup>
- Internal diameter of LAD or RCA is, according to the Japanese Ministry of Health Criteria.<sup>5</sup>
  - $> 3\text{mm}$ , if the patient is  $< 5$  years old
  - $> 4\text{mm}$ , if the patient is  $> 5$  years old
  - Segment diameter  $> 1.5$  times that of an adjacent normal segment
  - Lumen is irregular
- At least 3 of the following 6 criteria are met:
  - Perivascular brightness around the coronary artery lumen (see the discussion and illustration)
  - Coronary artery lumen lacks tapering from proximal to distal
  - Decreased left ventricular systolic function

- Mitral regurgitation
- Pericardial effusion
- Mild coronary dilatation ( $z = 2 \sim 2.5$ )

If echo is "positive," the patient should be treated. If the echo is negative, the patient's fever should be monitored. If fever persists, echo should be repeated and consultation should be obtained from someone who is experienced in KD.

**Step 4:** If CRP and ESR are not elevated, the patient should be followed daily without further testing. If fever resolves, then one should look for signs of typical desquamation. If typical peeling is observed, then echo should be obtained. In there is no peeling, the patient is dismissed from follow-up.

**Step 5:** If CRP and ESR are not elevated, but the patient continues to have fever for 2 more days, then the patient's clinical characteristics need to be reevaluated, as some KD patients

may show slower progression of systemic inflammation, and may still develop coronary artery complications. The importance of follow-up in such cases cannot be overemphasized.

#### Caveat about echo interpretation

**Use of Z score:** In the past, determination of coronary artery dilation relied on the criteria established by the Research Committee under the auspices of Japanese Ministry of Health and Welfare.<sup>5</sup> de Zorzi and coworkers compared the measurements of coronary arteries in the acute phase of KD to those of afebrile normal children, and concluded that some of the coronary arteries deemed normal on the basis of the Japanese criteria exceed 2 standard deviations above expected normal mean for the body surface area.<sup>4</sup> Z scores of the left main coronary artery, proximal LAD and proximal RCA are given in a graphic form in the published manuscript, and so the data will not be reproduced in this article. Although coronary Z scores have a higher sensitivity in detecting coronary dilatation

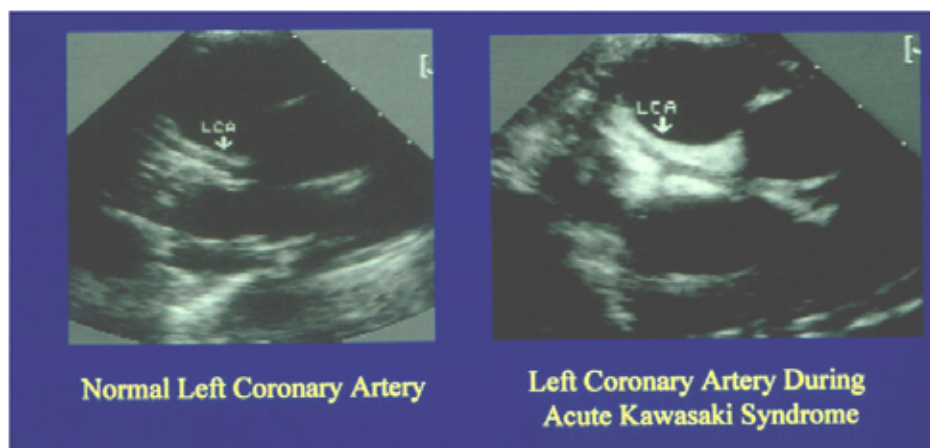


Figure 2. Examples of echocardiographic views of the left anterior descending artery in a modified parasternal long-axis view. Left panel: normal coronary artery with well-defined thin arterial walls. Right panel: perivascular echo brightness which obscures the arterial wall from the surrounding tissue.



california

**HEARTconnection**

[www.caheartconnection.org](http://www.caheartconnection.org) ■ Tel: 949-653-6421 ■ Patient handouts available

*A support and information network for those  
with congenital heart defects*



*"It has been 38 years since the original description of Kawasaki Disease (KD). KD is no longer a rare or uncommon disease. An average pediatric practitioner encounters this entity repeatedly in his/her practice."*

compared to the Japanese criteria, they do raise some concerns. As the control subjects were afebrile children rather than febrile controls, there is some possibility of low specificity. Also, since coronary arteries are only a few millimeters in diameter, any error in measurement may be amplified when one converts it to Z score. The sonographer must obtain the best and clearest possible image of the coronary artery and put the screen into zoom mode before undertaking measurements. Multiple measurements should be taken to ensure reproducibility.

**Perivascular echo brightness:** A normal coronary artery is characterized by a parallel thin (1-2 pixels wide) but intense echoes arising from the arterial wall, surrounded by less intense and non-uniform perivascular echoes (presumably from adventitia and perivascular fat tissues). Suzuki described appearance of enhanced perivascular brightness, which are uniformly intense perivascular echoes surrounding the lumen, appearing 5.4 ± 1.1 days from onset of fever. This perivascular echo brightness may be an echocardiographic reflection of edema and cellular infiltration due to arteritis. In 47.5% of cases, perivascu-

Table 1

**Classic Clinical Criteria**

- Fever for persisting at least 5 days \*
- Presence of at least 4 principal features:
  - ▲ Changes in extremities
    - ▲ Acute: Erythema of palms/soles; edema of hands, feet
    - ▲ Subacute: Periungual peeling of fingers, toes in week 2 and 3
  - ▲ Polymorphous exanthem
  - ▲ Bilateral bulbar conjunctival injection without exudate
  - ▲ Changes in lips and oral cavity: Erythema, lips cracking, strawberry tongue; diffuse injection of oral and pharyngeal mucosae
  - ▲ Cervical lymphadenopathy (>1.5 cm diameter), usually unilateral

Patients with at least 5 days and < 4 principal criteria may be diagnosed as KD if coronary artery abnormalities are detected by 2DE or angiography. Other possible diagnoses should be excluded.

\* In the presence of ≥4 principal criteria, Kawasaki disease diagnosis can be made on day 4 of illness. Experienced clinicians who have treated many Kawasaki disease patients may establish diagnosis before day 4.

lar brightness was accompanied by progressive dilatation of the lumen diameter (average 9.5 ± 2.7 days) and in 28.8% aneurysm formation (average 11 ± 1.9 days).<sup>6</sup> In our unpublished study, there was interobserver agreement of 85% among 5 cardiologists in detection of perivascular echo brightness (Figure 2).

**Current status of treatment:** A single infusion of IVIG 2 g/kg combined with aspirin remains the most effective treatment (based on level A evidence). However, > 10% of patients fail to defervesce with initial IVIG therapy. In such cases most experts recommend repeat administration of IVIG. As to the role of steroid as a rescue therapy, Hashino et al<sup>7</sup> in a small randomized study showed that those who received steroids had shorter duration of fever and lower medical costs than those who received an additional IVIG 1g/kg. No significant difference in coronary artery aneurysm incidence was noted

between the 2 groups. Corticosteroid as part of the primary therapy has undergone multicenter placebo-controlled randomized trial as a federally funded study, and its results are eagerly awaited.

Efficacy of other treatment modalities for patients who fail to respond to initial IVIG, including ulinastatin (neutrophil elastase inhibitor), abciximab (monoclonal antibody to Gp IIb/IIIa platelet receptor) and infliximab (monoclonal antibody to TNFα) are not yet proven.

**Summary:** The new AHA statement is a large document incorporating two previous documents on diagnosis and treatment and long-term management. It not only updates virtually all aspects of KD, but also attempts to raise the general awareness of incomplete (atypical) form of the disease. In this problematic subset of patients, the writing group departed from the traditional diagnostic guidelines, and crafted a

**NuMED**



[www.numed.on.ca](http://www.numed.on.ca)

**NuMED, Inc.**

Hallenweg 40, 5683 CT Best, The Netherlands

Tel: +31 (0)499 377388

Fax: +31 (0)499 377456

Manufacturer of angioplasty and valvuloplasty catheters, has a long standing commitment in meeting our customer's expectations by providing a high quality product. At NuMED, we see quality improvement as a continual process, aimed at satisfying these expectations and requirements at every stage.

***“Although the algorithm may seem complex at first glance, each component branch applicable to a particular patient’s situation should be quite useful in determining when to initiate treatment and when to obtain an echocardiogram.”***

complex algorithm, using patient’s physical characteristics, commonly used laboratory tests and echocardiographic findings, which include some findings other than coronary artery dimensions. Use of coronary Z score is recommended.

Although the algorithm may seem complex at first glance, each component branch applicable to a particular patient’s situation should be quite useful in determining when to initiate treatment and when to obtain an echocardiogram. It is our hope that this new diagnostic strategy will prove useful to many of the readers.

## References

1. Dajani AS, Taubert KA, Gerber MA, et al. *Diagnosis and therapy of Kawasaki disease in children*. Circulation 1996;94:1776-1780.
2. Newburger JW, Takahashi M, Gerber MA et al. *Diagnosis, treatment, and long-term management of Kawasaki disease: A statement for health professionals from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American heart Association*. Circulation 2004; 110:2747-2771, also Pediatrics 2004; 114:1708 – 1733.
3. Dajani AS, Taubert KA, Takahashi M,

et al. *Guidelines for long-term management of Patients with Kawasaki disease*. Circulation 1994;89:916-922.

4. deZorzi A, Colan SD, Gauvreau K, Baker AL, Sundel RP, Newburger JW. *Coronary artery dimensions may be misclassified as normal in Kawasaki disease*. J Pediatr 1998;133:254-258.

5. Research Committee on Kawasaki disease. *Report of Subcommittee on Standardization of Diagnostic Criteria and Reporting of Coronary Artery Lesions in Kawasaki Disease*. Tokyo, Japan: Ministry of Health and Welfare; 1984.

6. Suzuki A. *Standardized echocardiographic diagnosis of coronary artery lesions in Kawasaki disease*. Pediatrics Mook 1986; Suppl 1:160-170 [in Japanese].

7. Hashino K, Ishii M, Iemura M, Akagi T, Kato H. *Re-treatment for immune globulin-resistant Kawasaki disease: a comparative study of additional immune globulin and steroid pulse therapy*. Pediatr Int. 2001;43:211-217.

Email comments on this article to:  
JULYMT@CCT.bz

~CCT~



Masato Takahashi, MD  
Childrens Hospital Los Angeles  
Los Angeles, CA USA

Phone: +1.323.669.4634  
FAX: +1.323.669.7317

mtakahashi@chla.usc.edu

## USEFUL MEDICAL WEBSITES

**American College of Cardiology**  
[www.acc.org/](http://www.acc.org/)

**The Association for Europe Paediatric Cardiology**  
[www.aepc.org/home.htm](http://www.aepc.org/home.htm)

**BIOME**  
<http://biome.ac.uk/>

**BMJ Publishing Group Limited**  
<http://bmj.bmjournals.com/>

**British Heart Foundation**  
[www.bhf.org.uk/](http://www.bhf.org.uk/)

**Canadian Pediatric Cardiology Association**  
[www.cardioped-canada.org/cpa.asp](http://www.cardioped-canada.org/cpa.asp)

**CenterWatch Clinical Trials Listing Service**  
[www.centerwatch.com/](http://www.centerwatch.com/)

**CTSNet - The Cardiothoracic Surgery Network**  
[www.ctsnet.org/](http://www.ctsnet.org/)

**European Heart Network - EHN**  
[www.ehnheart.org/content/default.asp](http://www.ehnheart.org/content/default.asp)

**European Society of Cardiology**  
[www.escardio.org/](http://www.escardio.org/)

**FAC - Argentine Federation of Cardiology**  
<http://pcvc.sminter.com.ar/facen/indexeng.htm>

**PubMed**  
[www.PubMed.com](http://www.PubMed.com)

**South African Heart Association**  
[www.saheart.org/paeds.html](http://www.saheart.org/paeds.html)



## PICS/ENTICHS- 2005

Pediatric Interventional Cardiac Symposium

and Emerging New Technologies in Congenital Heart Surgery

SEPTEMBER 15-18, 2005 At the Hilton Buenos Aires Buenos Aires, Argentina

Immediately preceding the 4th World Congress of Pediatric Cardiology and Cardiac Surgery

[www.picsymposium.com](http://www.picsymposium.com)

## HIGHLIGHTS FROM THE 28TH ANNUAL SCIENTIFIC SESSION OF THE SOCIETY FOR CARDIOVASCULAR ANGIOGRAPHY AND INTERVENTIONS

By José A. Ettegui, MD

The 28th Annual Scientific Session of the Society for Cardiovascular Angiography and Interventions was held in Ponte Vedra Beach, Florida, from May 4-7, 2005. The Sawgrass Marriott Resort was the host venue and the facilities were spectacular. This year marked the 10th anniversary of the pediatric cardiology component of this event. This year's meeting was co-chaired by Robert Vincent, MD from Sibley Children's Heart Center, Atlanta and José Ettegui, MD (Pediatric Cardiovascular Center, University of Florida/Jacksonville. Bob Vincent (Co-chair) and Rick Henegar (SCAI) deserve special recognition for the hard work and

***"During the business meeting of the pediatric session, a consensus was reached regarding new directions for strategic planning. Top priority was given towards the development of training, credentialing and competency criteria for cardiologists performing congenital interventions in pediatric and adult patients."***

extra effort that translated into the success of this event. Pediatric session attendance was excellent, averaging over 80 people between physicians, allied health specialists and representatives from the industry.

The opening presentation for this meeting was delivered by Charles (Chuck) Mullins, MD (Houston, TX). He provided an eloquent historical review of transcatheter interventions from the early 1950's when Alvarez described use of a wire for perforating severely stenotic pulmonary valves to the current era. Chuck is a revered and respected father figure and pioneer in interventional pediatric cardiology. This was followed by Ziyad Hijazi's update on device closure of VSDs. The controversy section discussions centered on transcatheter vs. surgical treatment of coarctation of the aorta in children 2-6 years of age. Redmond Burke, MD (Miami, FL) made a strong case in support of surgical treatment (despite the 100% thoracotomy rate) and John Moore (Los Angeles, CA) showed beautiful images and convincing data on primary stent placement in these children. This was followed by a spirited discussion on management of the newborn with hypoplastic left heart syndrome. Emphasis was placed on the improving surgical results (Kirk Kanter, MD, Atlanta, GA) and the recognition of a steep learning curve for the inter-

***"The Abstracts session had 11 oral presentations that covered a wide variety of topics ranging from intraoperative collaboration between the cardiac surgeon and the interventionalist to the regional sharing of information through the MAGIC database."***

ventional approach (Mark Boucek, MD, Hollywood FL).

As predicted, the "I Blew It" session was a great hit. A variety of challenging and problematic patients were presented with no end to the mishaps that included coronary artery dissections, broken snares and embolized devices. This year's prize deservedly went to the cardiologist who presented chilling angiographic and echocardiographic clips of an embolized closure device. Fortunately for the patient and all else involved, there was a happy ending.

The Abstracts session had 11 oral presentations that covered a wide variety of topics ranging from intraoperative collaboration between the cardiac surgeon and the interventionalist to the regional sharing of information through the MAGIC database.

**PedHeart Suite**  
Congenital Heart Education

The most in-depth teaching materials  
for Congenital Heart Disease - anywhere!

From Patient Education to Senior Staff Review

[www.PedHeart.com](http://www.PedHeart.com)

Scientific  
Software  
Solutions



***"The opening presentation for this meeting was delivered by Charles (Chuck) Mullins, MD (Houston, TX)."***

During the business meeting of the pediatric session, a consensus was reached regarding new directions for strategic planning. Top priority was given towards the development of training, credentialing and competency criteria for cardiologists performing congenital interventions in pediatric and adult patients. In addition, major emphasis was placed on approaching the FDA with the concept of peer reviewed panels and objective performance criteria (OCP) for evaluation and approval of new devices and technologies. Finally, increased funding for fellow education and research in the form of abstract competitions and travel grants was discussed. These topics will be the subject of detailed discussions during a strategic planning meeting to be held in Washington, DC at the end of July.

The Friday morning sessions were memorable with a number of excellent presentations. Topics presented ranged from outcomes following valve perforation in infants with pulmonary atresia with intact septum (David Nykanen, Orlando, FL) through ductal stenting (John Cheatham, Columbus OH) to detailed features of stent technology (Frank Ng, San Diego, CA). Jim Huhta (St. Petersburg, FL) gave a fascinating presentation on fetal cardiac interventions that captured everyone's attention. The afternoon session (formerly combined adult/

pediatric) focused on device closure of patent foramen ovale (Tom Jones, Seattle, WA and Peter Block, Atlanta, GA) and wrapped up with percutaneous replacement of the aortic valve (Martin Leon, New York, NY) and repair of the mitral valve (Ted Feldman, Evanston, IL).

It was an honor and a pleasure hosting this event. The wealth and depth of information that was presented proves why the pediatric component of the annual scientific session of the SCAI has become established as one of the premier events for pediatric interventionalists. Mark your calendar for next years event to be held in Chicago from May 10-13, 2006. ([www.scai.org](http://www.scai.org)). See you there!

Email comments on this article to:  
JULYJAE@CCT.bz

~CCT~



*José A. Ettedgui, MD  
Glenn Chuck Professor of Pediatric Cardiology  
Chief, Division of Pediatric Cardiology  
University of Florida  
Jacksonville, FL USA  
Phone: +1.904.493.2290  
[jose.ettedgui@jax.ufl.edu](mailto:jose.ettedgui@jax.ufl.edu)*

#### **Submit an Article to Congenital Cardiology Today**

Send email with your name, title, organization, address, and email along with the title and a 50 word description to: [Articles@CCT.bz](mailto:Articles@CCT.bz)

#### **Dr. Barry F. Uretsky Installed as 2005–06 President of SCAI**

Barry F. Uretsky, MD, FSCAI, was installed as the 28th president of the Society for Cardiovascular Angiography and Interventions (SCAI) during the Society's Annual Scientific Sessions. Dr. Uretsky is Director of the Cardiovascular Catheterization Laboratory and Director of the Division of Cardiology at The University of Texas Medical Branch at Galveston.

Dr. Uretsky became a Fellow of SCAI in 1993 and has been active in the Society since that time. He chaired the SCAI Scientific Sessions in 2001 and launched the Society's newly created Development Committee the following year. These positions, in addition to his service as a Trustee since 2001 and as the Society's secretary in 2003–04, have given him important insights into the challenges that accompany a thriving medical society's growth.

Dr. Uretsky is known worldwide for his work on interventional cardiology techniques and his research on methods of cardioprotection. He has authored two textbooks, as well as more than 130 articles in peer-reviewed journals and 14 chapters appearing in edited volumes. Dr. Uretsky has served as the lead or co-investigator for nearly 50 research grants and contracts.

Dr. Uretsky graduated cum laude from Wesleyan University in Connecticut and earned his medical degree from Temple University School of Medicine in Philadelphia in 1972. He completed his internship, residency, and cardiology training in Boston, and then was a clinical instructor at Harvard Medical School for several years before moving to Pittsburgh. There, he co-directed cardiac catheterization services at Presbyterian University Hospital for 15 years and directed the heart-transplant service unit for six years. Dr. Uretsky also served as an associate professor of medicine at the University of Pittsburgh School of Medicine for much of that time. He moved to Texas in 1995 to head the cardiac catheterization lab and the interventional cardiology program at The University of Texas Medical Branch. Five years ago, he was named the University's Director of the Division of Cardiology.

## **Ask the Editorial Board**

Do you have a question about a procedure, technology or product that you would like to ask the Editorial Board's opinion? e-mail your question to: [ASK@CCT.bz](mailto:ASK@CCT.bz)

Selected questions and answers may be published in upcoming issues. Names will be withheld upon request.

*Nothing can compare to the look in the eyes of a child you've helped, especially when it involves matters of the heart. This is what cardiac care is all about. It's about turning medical challenges into extraordinary moments for everyone involved.*

# Start your day

doing what's good for your heart and hers.

At Children's National Heart Institute, you will thrive in an environment that is dedicated to family-centered care, where parents participate in their child's daily rounds and more. Most importantly, you will redefine the impact of pediatric cardiac care in an environment that is more focused and highly specialized than ever in such key areas as: inpatient and outpatient services, intensive care, surgery, catheterization and interventional therapies, electrophysiology, echocardiography, cardiac MRI, and fetal cardiology.

We're looking for the following physicians to be part of our exceptional team:

- **Pediatric Electrophysiology**
- **Cardiac Intensivist**
- **Heart Failure/Transplant**
- **Clinical Trials Director**

The professionals at Children's enjoy competitive salaries and benefits packages that are second to none.

Please contact us at: **202-884-2020** or e-mail your resume/CV to: **lmasters@cnmc.org**

EOE, M/F/D/V.



## MEDICAL NEWS, INFORMATION &amp; PRODUCTS

**PICS and ENTICHS Announces....**

For the first time the creation of "Breakout Sessions" designed specifically to address issues related to the catheterization laboratory and OR environment will be added to the PICS & ENTICHS 2005 program in Buenos Aires, September 15-18, 2005.

These sessions will address the role of nurses and techs in such places, and will emphasize the importance of such professionals in management of neonates/infants and children with critical heart disease. The sessions will be given by leaders in this field, including Drs. Gilbert Wernovsky, John P. Cheatham, Julie Vincent and Nurse Practitioners, Erika Speier and Sharon Hill.

Bring one of your nurses or techs with you to Buenos Aires to learn the tricks of the trade. For registration and more information: [www.picsymposium.com](http://www.picsymposium.com)

**Heart Failure Patients do Better When Treated According to Guidelines, But Too Many Doctors Still Fail to Prescribe the Best Treatments**

Heart failure patients who are treated in accordance with established European guidelines do better than patients who are not, yet many doctors are still not adhering to the guidelines, according to pioneering research published today (Tuesday 3 May) in Europe's leading cardiology journal, the European Heart Journal.

In the first large European study to look at the effect of prescribing practices on outcome in heart failure outpatients outside of a clinical trial<sup>1</sup>, Professor Michel Komajda and colleagues investigated the way that 1,410 patients with mild to moderate heart failure were treated by 150 randomly selected cardiologists or cardiology departments in six European countries (France, Germany, Italy, The Netherlands, Spain and the UK). They measured how closely the patients' treatment adhered to the guidelines issued by the European Society of Cardiology (ESC) for the use of the five most commonly used cardiac drugs: ACE-inhibitors, beta-blockers, spironolactone, diuretics and cardiac glycosides.

Prof Komajda, professor of cardiology at the Pitie-Salpetriere Hospital, Paris, France, and a specialist in heart failure, said, "We found that where doctors had treated their patients in accordance with the ESC guidelines, fewer patients had to be referred to the hospital due to deterioration of their heart failure or for cardiovascular symptoms, and there was a longer time before patients had to be readmitted to hospital because of their symptoms."

"However, the study showed that only 60% of patients were treated according to the ESC guidelines with ACE-inhibitors<sup>2</sup>, beta-blockers or spironolactone<sup>3</sup> – the three cardiac drugs for which there is the strongest evidence of benefit – and only 63% of patients were treated ac-

cording to the guidelines for these three drugs plus the two other commonly used drugs, cardiac glycosides and diuretics.

"This means there is a high proportion of patients who are not receiving the best possible treatment for their condition, and who suffer worsening symptoms and even death as a result."

Patients in the MAHLER study<sup>4</sup> were aged 40 or over, with an average age of nearly 69. They were followed up for six months. The researchers found that while adherence to diagnosis guidelines was high at 74%, adherence to treatment guidelines was much lower, with large variations between the five different treatments; 85% of patients who needed them were prescribed ACE-inhibitors, 58% were given beta-blockers, 83% a diuretic agent, 52% a cardiac glycoside, and only 36% spironolactone.

When the researchers looked at the impact on outcome of the "big three" drugs (ACE-inhibitors, beta-blockers and spironolactone) they found that amongst patients who were treated with perfect adherence to the guidelines 6.7% and 11.2% were admitted to hospital with chronic heart failure (CHF) or a worsening of their cardiovascular (CV) symptoms respectively; this compared with figures of 9.7% and 15.9% respectively for moderate adherence and 14.7% and 20.6% for low adherence. These outcomes were independent of the severity of the disease, previous hospitalizations for CHF, or the presence of high blood pressure or diabetes.

The researchers found there was plenty of room for improvement in the treatment of patients with heart failure. "Prescription of beta-blockers was observed in only half of the patients who should be receiving them, according to ESC guidelines. In contrast, a substantial proportion of our patients received cardiac glycosides despite the fact that this treatment is only recommended for symptomatic improvement. Overall, we found a high level of compliance to guidelines for ACE-inhibitors or diuretics, but a rather low level for beta-blockers, suggesting that there is still room for improving practice in Europe and that there is a need to develop training programmes to improve the quality of care for cardiac patients," said Prof Komajda.

**Reference:**

1. Adherence to guidelines is a predictor of outcome in chronic heart failure: the MAHLER survey. European Heart Journal. doi:10.1093/eurheartj/ehi251
2. ACE-inhibitors are angiotensin-converting enzyme inhibitors.
3. Spironolactone is a type of diuretic.
4. MAHLER stands for Medical mAnagement of chronic Heart failure in Europe and its Related costs.

For more information on this and related articles visit ESC's website at: [www.escardio.org](http://www.escardio.org).

**AMPLATZER®** Septal Occluder



Simple. Reliable. Repositionable.

For more information visit: [www.amplatzer.com](http://www.amplatzer.com) or call: 763-513-9227  
Toll free in the US: 888-546-4407





# PAEDIATRIC CARDIOLOGY LOSES A PIONEER: ROBERT M. FREEDOM MD, FRCP(C), FACC, O ONT 1941-2005

By Joel A. Kirsh, MD; Lee N. Benson, MD; John Pires

Dr. Robert M. Freedom passed away on Saturday May 7, 2005, in Nova Scotia, Canada.

Dr. Freedom was known around the world as one of the fathers of Paediatric Cardiology, and served three five-year terms as head of the Division of Cardiology at the Hospital for Sick Children in



Dr. Robert Mark Freedom, MD, FRCP(C), FACC, O Ont

Toronto. He was internationally recognized for his clinical skills, the training of academic cardiologists from around the world, and as a prolific author of original clinical research and textbooks, several of which are considered classics in the field.

Born and educated in the United States, Dr. Freedom moved to Toronto in 1974 to join the Division of Cardiology at Sick Kids, after two years on staff at Johns Hopkins in his hometown of Baltimore. He began his first term as head of Cardiology in 1986. At the time of his retirement in 2000, Dr. Freedom was Emeritus Professor of Paediatrics and Pathol-

ogy at the University of Toronto. Amongst the many honours and awards during his career were the PAIRO (Professional Association of Interns and Residents of Ontario) Award for Teaching Excellence, and the Council Award of the College of Physicians and Surgeons of Ontario. Dr. Freedom was named to the Order of Ontario in 2000, the province's highest civilian honour. Last

"My training with Bob at Sick Kids was so important and valuable. A great scholar, teacher and man. Did you call the referring doctor with an update on his/her patient.... Yes Bob, I left a message.....Call the doctor and speak to them personally....Yes Bob, I mean Dr. Freedom.

Communication with the child, the family and the referring doctor were what he taught me to do to this day. Thank you Bob. I pray that you will enjoy meeting the Pope again up there. You know the guy in the white clothes standing next to you Bob in that picture you were so proud of—bigger than life."

~ Roland Beaulieu, MD

"I remember Bob from his days at UCLA way back when. Bob's career in pediatric cardiology has been nothing short of spectacular. He had the unique gift of finding insight into pathophysiology by studying cardiac anatomy. He has done a great deal in enhancing initial palliation of many a difficult congenital heart disease."

~ Masato Takahashi, MD

"Our association as residents and fellows at CHMC, Boston together from 1967-1972 is a close one. I will remember those good old days and you for all the years to come"

~ Charlee Phornphutkul, MD

"We all miss Big Bob dearly. But his enormous contribution to the Pediatric Cardiology World will live on many years to come."

~ Sang Park, MD

**"Dr. Freedom was known around the world as one of the fathers of Paediatric Cardiology, and served three five-year terms as head of the Division of Cardiology at the Hospital for Sick Children in Toronto."**

month, Dr. Freedom was recognized by the journal Cardiology in the Young, ([www.greenwich-medical.co.uk/](http://www.greenwich-medical.co.uk/)) joining the Paediatric Cardiology Hall of Fame. A seven-page tribute, including a recent interview and list of fellows trained by Dr. Freedom, appears in the April issue of Cardiology in the Young. (Benson LN, Anderson RH. Robert Mark Freedom MD, FRCP(C), FACC, O Ont. Cardiology in the Young 15(2):206-12, April 2005).

As per Dr. Freedom's wishes, no funeral was held, and a tribute was held on Sunday May 29th, at the conclusion of the first day of the Toronto Symposium in Congenital Heart Disease. Dr. Freedom's personal website, [www.granvilleferry.com](http://www.granvilleferry.com), has photos and a guestbook for visitors.

Email comments on this article to:  
JULYRMF@CCT.bz

## Editor's Note

Between July 1970 and April 2005, Dr. Freedom authored or co-authored over 360 articles in a number of major medical publications including The American Journal of Cardiology, Circulation, Cardiology in the Young, Journal of Pediatrics, Radiology, Johns Hopkins Medical Journal, The British Heart Journal and The European Journal of Cardiology to mention just a few. To see this impressive list, visit Entrez PubMed ([www.PubMed.com](http://www.PubMed.com)), type "Freedom RM" in the Search box.

# CONGENITAL CARDIOLOGY TODAY®

RELIABLE INFORMATION IN CONGENITAL CARDIOLOGY™

## INDEX OF SPONSORS

AGA Medical Corporation - pages 3, 14  
[www.amplatzer.com/international/index.html](http://www.amplatzer.com/international/index.html)

California Heart Connection— page 8  
[www.caheartconnection.org](http://www.caheartconnection.org)

The Fourth World Congress of Pediatric Cardiology and Cardiac Surgery - page 6  
[www.pccs.com.ar](http://www.pccs.com.ar)

NuMed - pages 5, 9  
[www.numed.on.ca](http://www.numed.on.ca)

PICS/ENTICHS- 2005 (Pediatric Interventional Cardiac Symposium and Emerging New Technologies in Congenital Heart Surgery) - pages 2, 10  
[www.picsymposium.com](http://www.picsymposium.com)

Scientific Software Solutions - pages 7, 11  
[www.PedCath.com](http://www.PedCath.com)

## RECRUITMENT ADVERTISEMENT

Childrens National Medical Center - page 12  
[www.dccchildrens.com](http://www.dccchildrens.com)

## UPCOMING ARTICLES IN FUTURE ISSUES

Pfm Device Closes Perimembranous VSDs *by John W. Moore, MD*

Estimates of the Burden of Congenital Heart Disease in the Developing World *by R. Krishna Kumar, MD*

Highlights from Update on Cardiac Care in Neonates, Children, and Young Adults *by Anthony C. Chang, MD*

Dysphagia Lusoria in a Neonate *by Christopher J Knott-Craig, MD*

Cardiac Progenitor Cells and Regenerative Medicine - a Cautionary Tale *by Paul J. Grueber, MD and Karl-Ludwig Laugwitz, MD*

Mechanical Circulatory Assist Devices in Children with Therapy-refractory Heart Failure: a Review *by Felix Berger, MD and Brigitte Stiller, MD*

The Global Burden of Congenital Heart Disease *by Balu Vaidyanathan MD, DM and R. Krishna Kumar MD, DM, FACC*

Highlights from PEDIRHYTHM 2 *by Alpay Celiker, MD*

Highlights from PICS / ENTICHS—2005 *by Ziyad Hijazi*

12th World Congress on Heart Disease *by Asher Kimchi, MD*

© 2005 by Congenital Cardiology Today (ISSN 1554-7787-Print; ISSN 1554-049-online). All rights reserved. Photocopying, reproduction or quotation either in full or in part is strictly prohibited without the written permission of the publisher. For permission, send an e-mail to: [Permission@CCT.bz](mailto:Permission@CCT.bz). Statements or opinions expressed in Congenital Cardiology Today reflect the views of the authors and are not necessarily the views of Congenital Cardiology Today. Postmaster: Send address changes to Congenital Cardiology Today, 9008 Copenhagen Dr., Suite M, Potomac, MD 20854 USA

### Article Submission

[Article@CCT.bz](mailto:Article@CCT.bz)

### Information

[Info@CCT.bz](mailto:Info@CCT.bz)

### Medical Editor/Medical Editorial Board

John W. Moore, MD, MPH, FACC  
[jwmoore@mednet.ucla.edu](mailto:jwmoore@mednet.ucla.edu)

### Contact the Editorial Board

See page 2 for a complete list of Editorial Board members and how to contact them.

### New Product Submission

[PR@CCT.bz](mailto:PR@CCT.bz)

### Article Reprint Information

[Reprint@CCT.bz](mailto:Reprint@CCT.bz)

### Web Site Information

[Web@CCT.bz](mailto:Web@CCT.bz)

### Symposium Submission

[Symposium@CCT.bz](mailto:Symposium@CCT.bz)

### Medical Website Submission

[URL@CCT.bz](mailto:URL@CCT.bz)

### How to Reference CCT

[Reference@CCT.bz](mailto:Reference@CCT.bz)

### FREE Subscription

Congenital Cardiology Today® is published monthly and FREE to qualified professionals worldwide in congenital and pediatric cardiology, and related fields.

Print subscriptions are available in the U.S. and Canada for the North American edition.

Electronic subscriptions (in Adobe PDF format) are available in the International edition. To subscribe to either the International or North American edition, send an email with your name, title, organization, phone, address and e-mail to: [Subs-Int@CCT.bz](mailto:Subs-Int@CCT.bz). Or fax your request to +1.240.465.0692

### Sponsorship Production

[Production@CCT.bz](mailto:Production@CCT.bz)

### Letters to the Editor

[Letters@CCT.bz](mailto:Letters@CCT.bz)

### Sales & Marketing

[Sales@CCT.bz](mailto:Sales@CCT.bz)

### Recruitment/Fellowship Advertising

[Recruitment@CCT.bz](mailto:Recruitment@CCT.bz)

### Publishing Management

Tony Carlson, Founder  
Tel: +1.301.279.2005  
Fax: +1.240.465.0692  
[TonyC@CCT.bz](mailto:TonyC@CCT.bz)

Richard Koulbanis, Editor & Publisher  
[RichardK@CCT.bz](mailto:RichardK@CCT.bz)

Virginia Dematatis, Editorial Consultant  
[VirginiaD@CCT.bz](mailto:VirginiaD@CCT.bz)

Loraine Watts, Editorial Assistant

Caryl Cornell, Editorial Assistant

### Sales Office

CONGENITAL CARDIOLOGY TODAY  
9008 Copenhagen Drive, Suite M  
Potomac, MD 20854 USA  
Tel: +1.301.279.2005  
Fax: +1.240.465.0692

### Editorial Office

CONGENITAL CARDIOLOGY TODAY  
19509 Pine Cone Court, Suite 100  
Gaithersburg, MD 20879 USA  
[Editorial@CCT.bz](mailto:Editorial@CCT.bz)

[www.CongenitalCardiologyToday.com](http://www.CongenitalCardiologyToday.com)