# CONGENITAL CARDIOLOGY TODAY

News and Information for Pediatric and Congenital Cardiovascular Physicians and Surgeons

Vol. 4 / Issue 11
November 2006
International Edition

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## TETRALOGY OF FALLOT (TOF) WITH ANOMALOUS LEFT CORONARY ARTERY FROM PULMONARY ARTERY (ALCAPA) AS A RARE PRESENTATION OF NEONATAL HEART FAILURE


*By Mashail AL-Obaidan, MD; Zuhair AL-Halees, MD, RCSC, FACS, FACC, FACCP*



### Abstract

Coronary artery anomalies are common among patients with tetralogy of Fallot, however, the origin of the left coronary artery from the main pulmonary artery in a patient with TOF is rarely seen and not yet reported in the literature. The patient reported here presented at three weeks of age with impaired cardiac function and was found to have this rare anomaly.


### Introduction

Tetralogy of Fallot is considered one of the most common cyanotic congenital heart defects. It is characterized by VSD, hypoplastic stenotic pulmonary valve and main pulmonary artery, hypoplastic outlet septum (infundibulum), and overriding of the aorta.

Associated coronary abnormalities are quite common, including RCA from LCA, LAD from RCA with LAD crossing the RV outflow tract, and single coronary ostium. There is also a single case report of RCA arising from the MPA in TOF associated with Pulmonary Atresia.

The patient in this case report had TOF with ALCAPA. This specific combination of abnormalities has not to our knowledge, been previously reported. We report the initial presentation and management of the patient, and review the literature.

### Case report

A 1.5 month old boy weighing 3.2 Kg was referred from the periphery with congestive heart failure. He had two previous admissions due to chest infection and cardiomegally. There was a history of low grade fever, cough and shortness of breath ten days prior to the first admission associated with blue discoloration of the lips. After complete blood work and screening for sepsis, he was treated for seven days with antibiotics and was started on anti-failure medications (digoxin, lasix, and captopril). He was referred to our institute because of a heart murmur to rule out congenital heart disease.

> ***"Three months after hospital discharge, the patient was thriving and had no cyanosis. He was taking anti-failure medications. Echocardiography revealed further improvement in left ventricular function with ejection fraction of 45%."***

On examination, he had no dysmorphic features. He was pale and tachypneic, had nasal flaring, and his saturation in room air was 84%. His lung examination was not remarkable, but his abdominal examination revealed a liver edge 4 centimeters below the costal margin. His cardiovascular examination showed normal S1, single S2, and a systolic ejection murmur most prominent at the left upperstern al border. The remainder of his examination was normal for his age.

Laboratory investigation showed: normal CBC with Hemoglobin of 14 gm/dl. Serum electrolytes and liver function tests were

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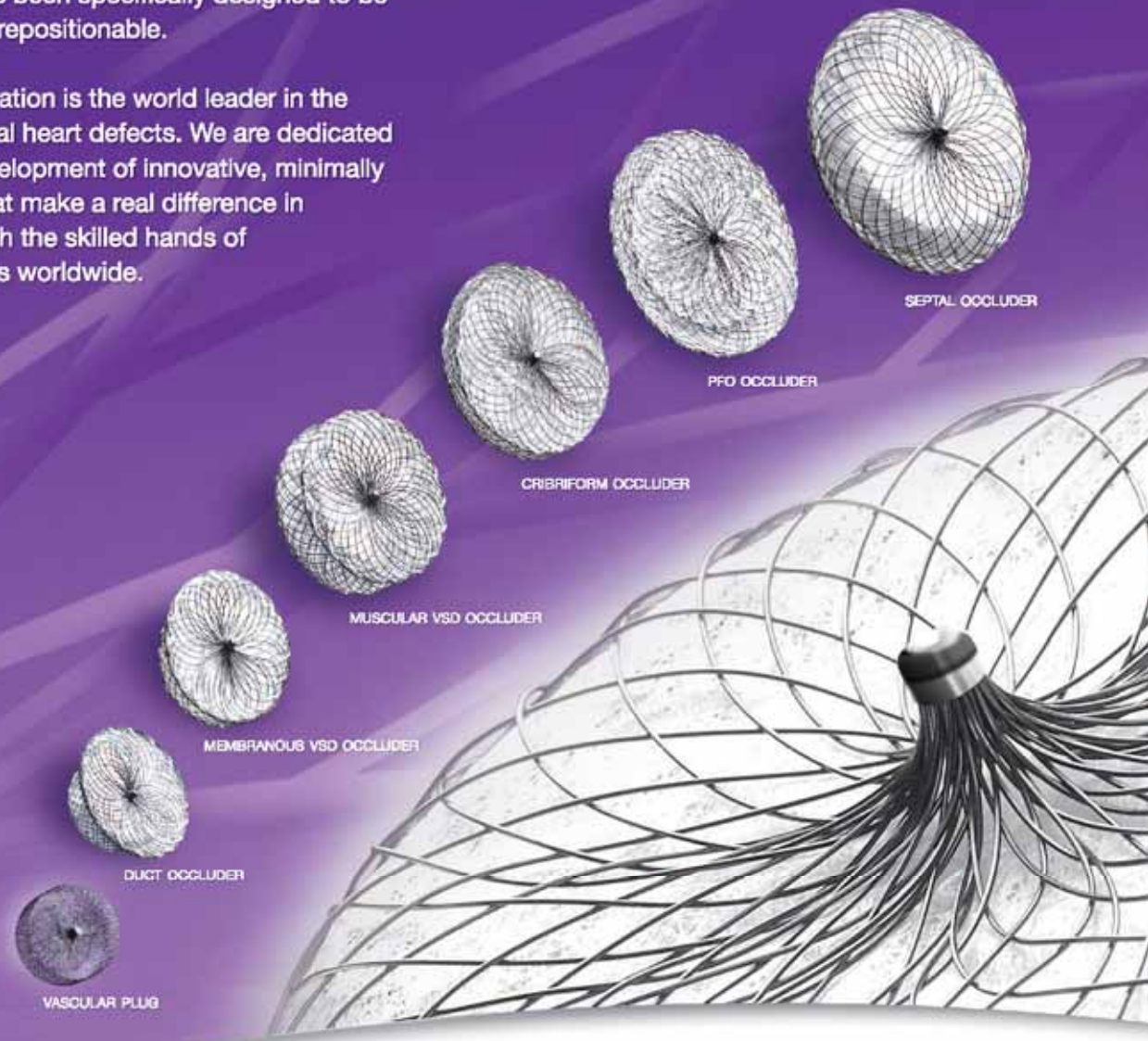


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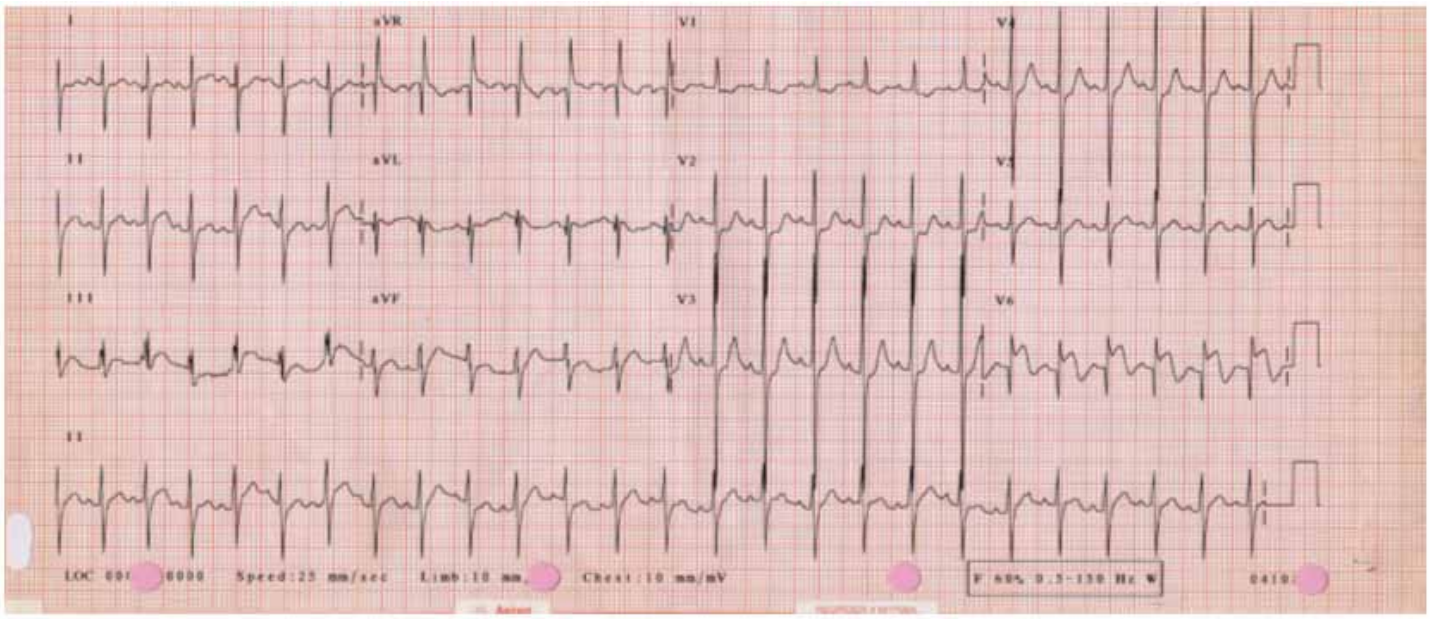

*Figure 1. ECG of the patient showing deep Q wave, ST elevation and RVH.*

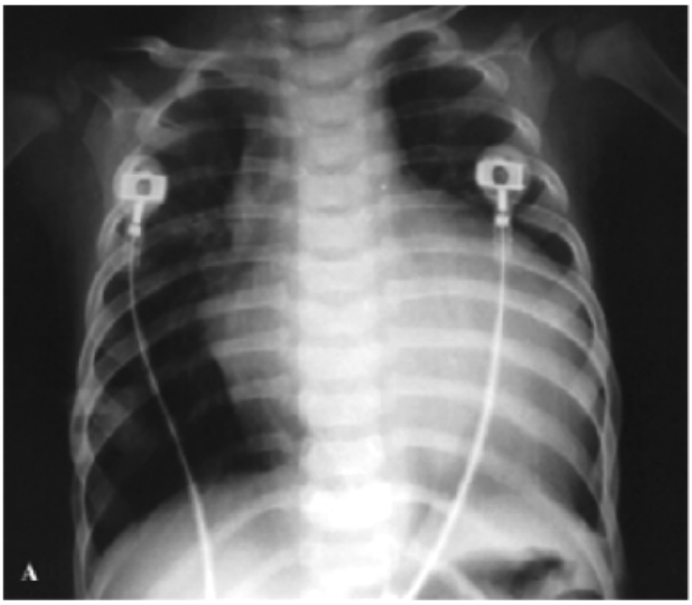

*Figure 2. Chest X-ray of the patient which revealed cardiomegally and oligaemic lung fields.*

within normal limits. Urine analysis was negative. Blood and urine cultures were negative. Serum virology studies were also negative. Serum carnitine was normal. LDH was 564 (elevated).

**ECG:** S/R, RAD, RVH, deep Q wave especially in lead V6, ST elevation (Figure 1).

**Chest X-ray:** cardiomegally, oligaemic lung field (Figure 2).

**Echocardiography:** Showed TOF with malaligned VSD, overriding aorta, infundibular and pulmonary valve stenosis with maximum velocity of 4.4 m/s (peak gradient 76 mm Hg), dilated RCA, myocardial coronary collaterals, LCA from MPA, markedly impaired left ventricular systolic function (ejection fraction of 15%, shortening fraction < 10%), PFO, mild mitral valve regurgitation (Figures 3-6).

**Cardiac Catheterization:** Proved the presence of TOF and ALCAPA (Figure 7-9).

**Hospital Course**

After completing these investigations and confirming the diagnosis, the patient was scheduled for surgical repair. Surgery included repair of TOF using the transatrial approach with VSD closure and resection of the infundibular stenosis and pulmonary valvotomy. Direct reimplantation of the LCA to the ascending aorta was also performed. Cross-clamp time was 90 minutes, and bypass time was 140 minutes. The patient's chest was left opened for 36 hours, and the patient required standard inotropes

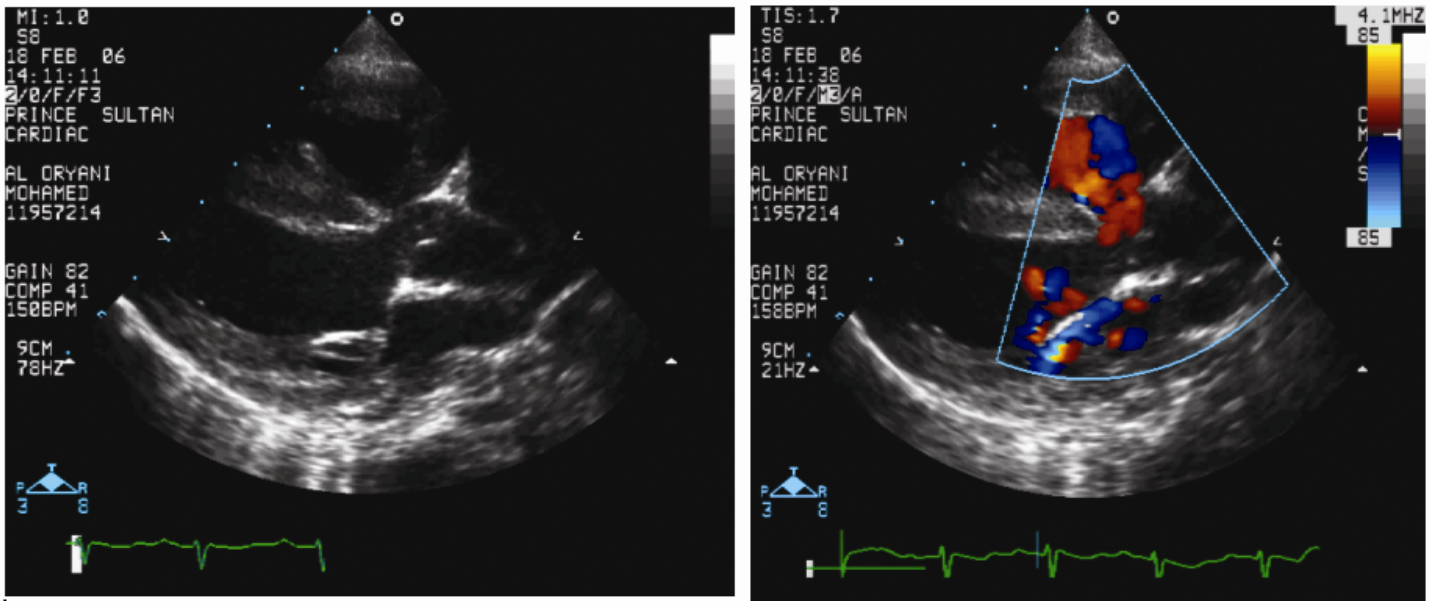


*Figure 3. Echocardiography of the patient showed malalinged VSD & overriding of aorta.*

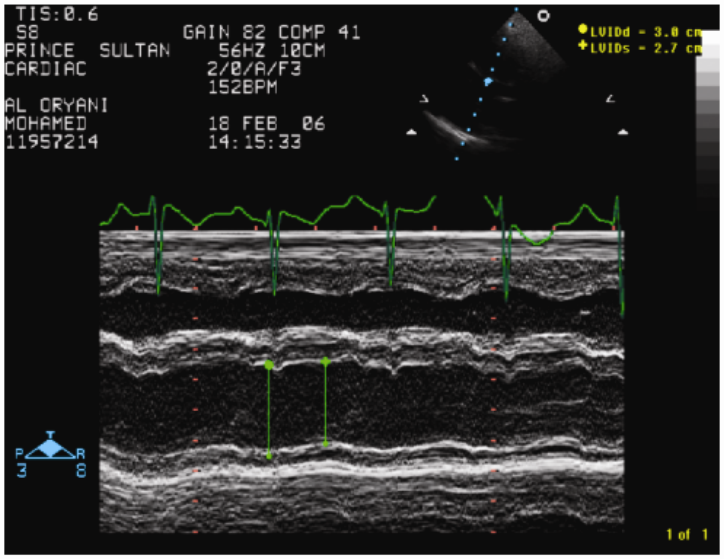


*Figure 4. M-mode revealed impaired LV systolic function.*

(dopamine, dobutamine, and milrinone). The patient remained in sinus rhythm, and his ECG normalized on the 3rd post-operative day. He required ventilation for 6 days, and stayed for an additional 11 days in the hospital. Echocardiography performed seven days after surgery revealed no residual VSD and no evidence of residual pulmonary stenosis. Left ventricular function was improved with ejection fraction of 30%.

### Follow Up

Three months after hospital discharge, the patient was thriving and had no cyanosis. He was taking anti-failure medications. Echocardiography revealed further improvement in left ventricular function with ejection fraction of 45%.

### Discussion

Coronary artery anomalies are often associated with tetralogy of Fallot. The left anterior descending coronary artery arising from the right coronary artery and crossing the RVOT is the commonest of them with reported incidence of 2-9% (1,2,3).

Additional anomalies include single coronary ostium, origin of the RCA from the LCA (4), a significant conal branch from the RCA (or a dual LAD), paired anterior descending arteries originating from left coronary artery and right coronary artery as well as RCA from LAD (5).

Anomalous origin of the LCA from the pulmonary trunk (ALCAPA) is a well described, although rare, congenital

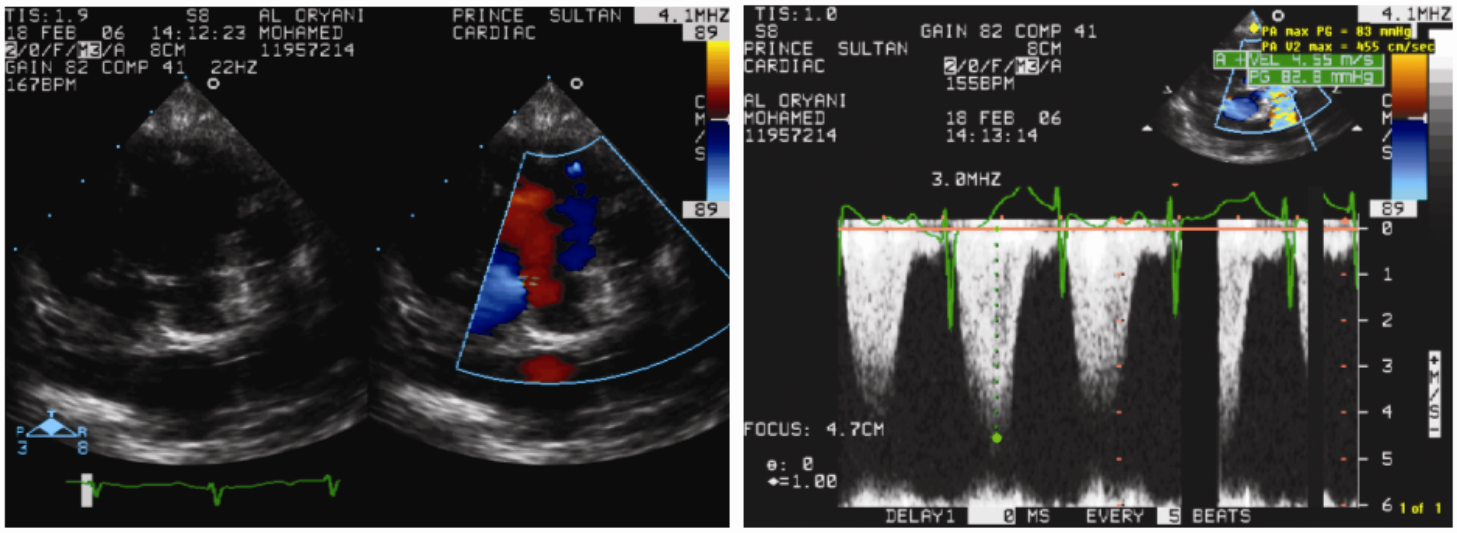


*Figure 5. parasternal short axis view showing malalinged VSD with infundibular and pulmonic valve stenosis v max @ 4.5 m/s.*

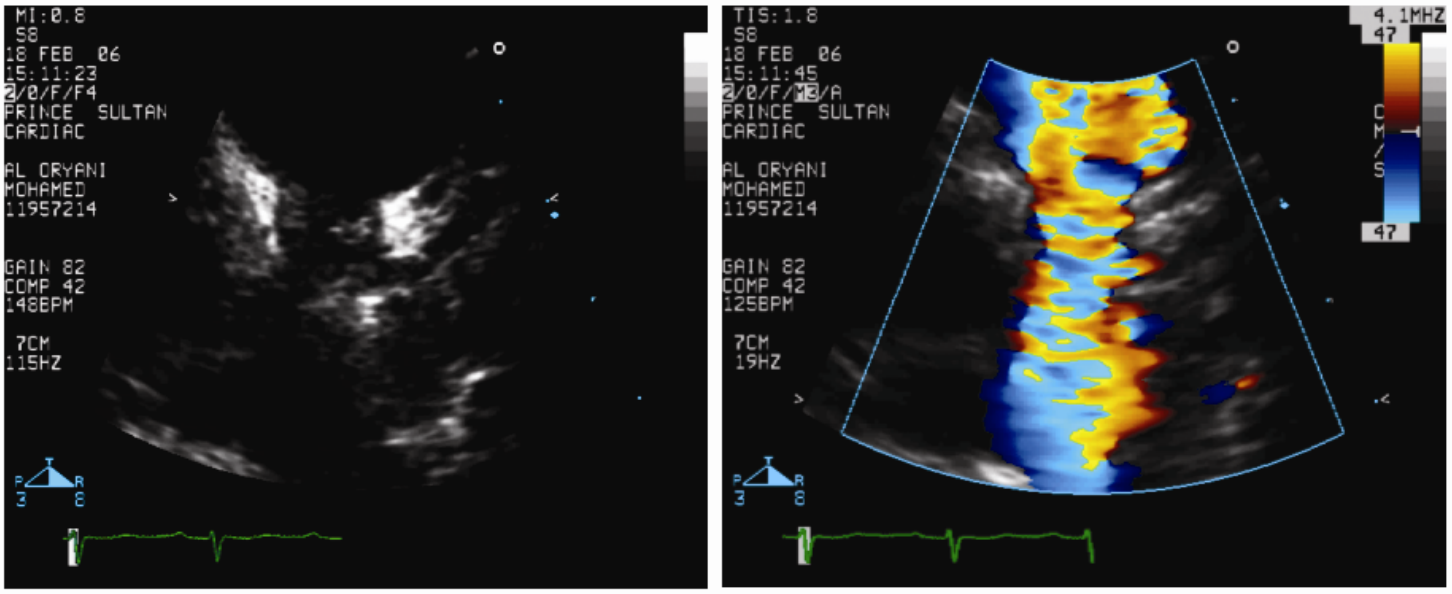


*Figure 6. 2-D and color-Doppler showing pulmonic valve stenosis with LCA arising from the pulmonary artery.*

anomaly in humans, with an incidence as an isolated anomaly of about one case for each 300,000 live birth. It represents 0.5% of the total cases of congenital heart disease. This condition is often characterized by a single origin of the whole left coronary artery from the posterolateral wall of the pulmonary trunk. The majority of patients present in infancy after 2 months of age (the time of normalizing pulmonary vascular resistance) with a picture of congestive heart failure and angina. Almost all die in the first year of life if they are not treated (6,7). A few reports describe older individuals with the disease (7).

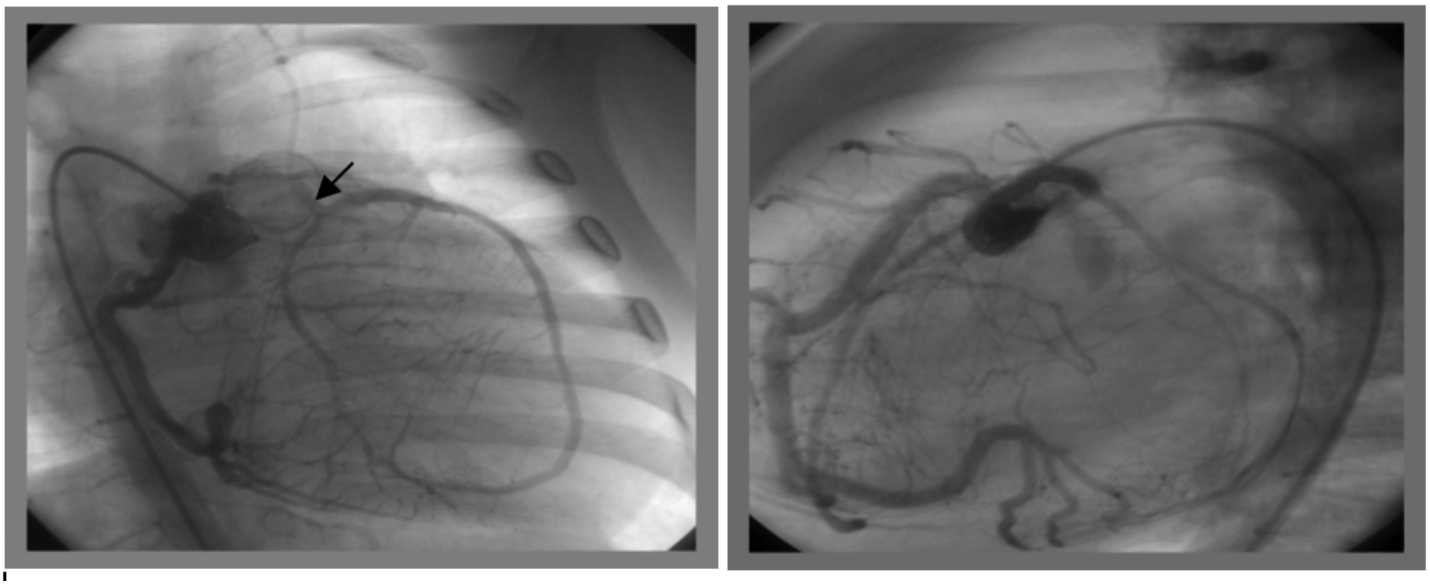

*Figure 7. Aortogram (RAO and LAO) revealed RCA filling from aorta & LCA filling from RCA through collaterals and Rt aortic arch (arrow indicate LCA).*

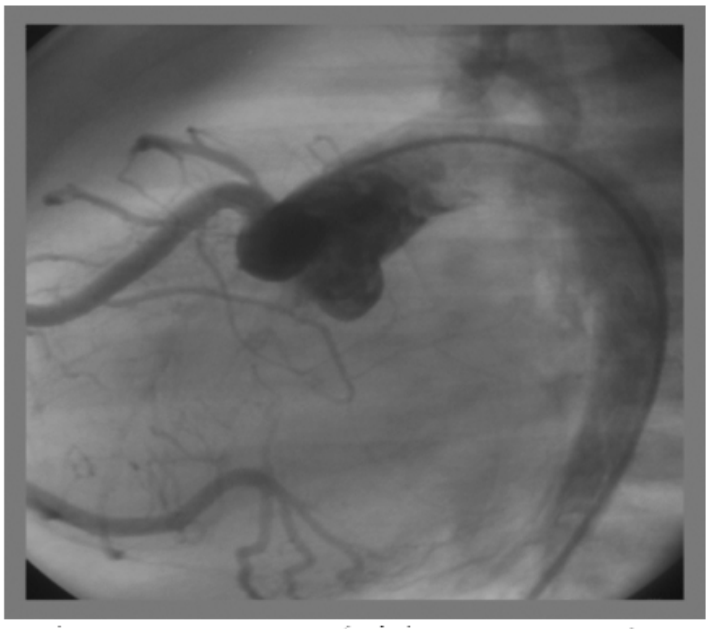

*Figure 8. Aortogram revealed dilated RCA.*

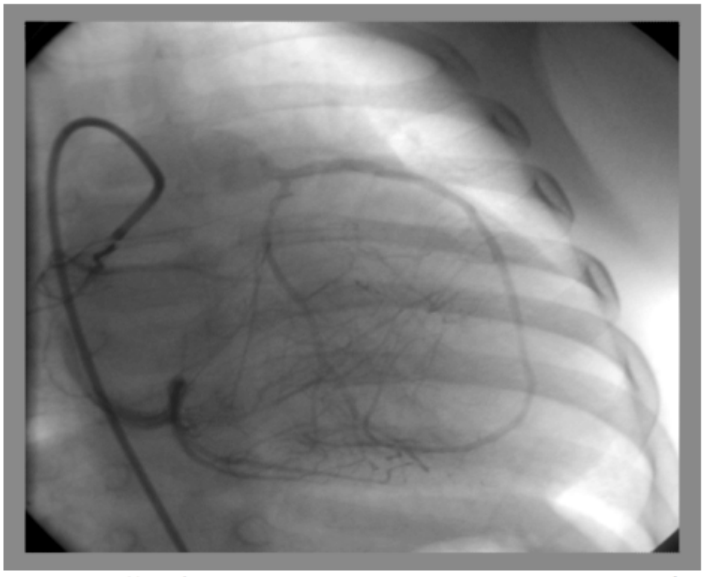

*Figure 9. Selective RCA angiogram revealed filling of LCA from collaterals arising from the RCA.*

The association of tetralogy of Fallot and anomalous left coronary artery from the main pulmonary artery has not been previously reported. There are scattered reports of TOF with RCA origin from the MPA (8), and TOF and PA with MPA arising from the RCA or LCA wherein the coronary arteries arise from the aorta and are the sole blood supply to the pulmonary artery (9,10). It is also worth mentioning that there are a few reported cases of coronary to pulmonary artery collaterals in which the coronary arteries arise normally from the aorta in patients with TOF and PA (11).

The clinical presentation of our patients was earlier than the typical presentation of patients with isolated ALCAPA. Our patient presented in the neonatal period with congestive heart failure and markedly impaired left ventricular function.

The earlier presentation of congestive heart failure in patients with TOF and ALCAPA may be explained by lower pulmonary vascular resistance in the neonate born with pulmonary stenosis and infundibular stenosis. In contrast patients with isolated ALCAPA may have a more delayed fall in pulmonary vascular resistance. This delays the steal phenomenon, the ischemic insult to the left ventricle and the development of congestive heart failure.

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***"Coronary artery anomalies are common among patients with tetralogy of Fallot, however, the origin of the left coronary artery from the main pulmonary artery in a patient with TOF is rarely seen and not yet reported in the literature."***

Our patient underwent successful total repair of tetralogy of Fallot using a transatrial approach with VSD closure and resection of the infundibular stenosis and pulmonary valvotomy, as well as direct reimplantation of the anomalous left coronary artery to the ascending aorta (12).

The patient had a favorable immediate and early result as well as in the first follow up period with improvement of the ventricular function.

From this case, we learned that any type of coronary artery anomaly can present in conjunction with tetralogy of Fallot, including ALCAPA. Furthermore the combination of TOF and ALCAPA may result in an early and devastating patient presentation, requiring prompt diagnosis and treatment.

**Acknowledgement**

To Mrs. Farida AL-Hazmi the senior echocardiographer for her grateful help in retrieving the best echo, images.

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~CCT~

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# HIGHLIGHTS FROM PICS 2006, LAS VEGAS, NEVADA

*By Ziyad M. Hijazi, MD*

With just about 1000 attendees from 60 countries in addition to the USA, PICS (Pediatric and Adult Interventional Therapies for Congenital and Valvular Heart Disease) was a huge success. 60% of the attendees were from the US, and 40% from abroad. I was told many of our international colleagues could not get a visa to the US!

For the first time, this year's meeting included combined and breakout pediatric and adult sessions for interventional therapy of congenital and structural heart disease. Live cases from 13 sites were transmitted via satellites to the venue at the Bellagio Hotel. The response we received so far is overwhelming with excellent feedback about the educational contents and the venue.

PICS started Sunday Sep. 10th, 2006 at 9:00 AM with an industry-sponsored symposium (AGA Medical Corp. Annual Meeting). This was very well attended. In this symposium, the Sim Suite was featured for PFO and ASD closure. Dr. Cheatham and Dr. Ruiz went over the cases with the attendees to demonstrate the protocol and closure steps. The addition of the Sim Suite to the tools available to the interventional cardiologists will greatly enhance the training and certification of physicians with the use of the Amplatzer devices.


The afternoon was marked by an exciting workshop on ASD's and PFO's. In this workshop, the majority of devices/techniques used (investigational and approved) for ASD and PFO closure were discussed. A total of 14 speakers discussed various devices and topics related to PFO and ASD closure. In addition to the devices used for closure, the use of ICE was discussed as well as the use of TCD to detect right to left shunt.

At the same time as the workshop, there were simultaneous sessions of oral abstract presentations. A total of 30 abstracts were selected out of 145 for oral presentations in congenital and valvular heart disease. This was followed by "meet the expert sessions." Again, this was a combined session between adult and pediatric cardiologists to discuss difficult cases that were presented by the attendees.

At the end of the day, a welcome reception was held in the exhibit area that held more than 30 exhibits representing different manufacturers of devices, catheters, etc.

The following day, Monday, September 11th was a combined day between the adult and pediatric cardiologists. The symposium was officially inaugurated and the activities started with live cases from Milan, followed by live cases from Frankfurt, Sao Paulo and Toronto. In these cases the operators discussed various cases of congenital heart disease in adult patients. From Milan, Drs. Mario Carminati and Gianfranco Butera operated together and demonstrated excellent cases; from Frankfurt, Dr. Horst Sievert demonstrated two cases of PFO closure using different devices; Dr. Carlos Pedra, from Sao Paulo, demonstrated three excellent cases of various interventions and Drs. Lee Benson and Eric Horlick from Toronto demonstrated excellent two cases including a percutaneous pulmonary valve implantation.

The first lecture of the day was by Howard Herrmann and David Nykanen about the technique of trans-septal puncture in adults and children. This was followed by an excellent lecture by Bill Hellenbrand on coarctation stenting.

During the entire day, nine cases were transmitted live from the above mentioned centers, which demonstrated the latest techniques for adults with congenital heart disease. Further, ten

lectures from the course faculty were given addressing various topics including as mentioned above, trans-septal puncture techniques, coarctation stenting and a series of talks about the right ventricle outflow tract (given by Charlie Kleinman, Dianna Bardo, James Wilkinson, Kevin Walsh, Philipp Bonhoeffer and Ziyad Hijazi)! Finally, Drs. Ted Feldman and Mike Mullen gave a nice presentation on how to develop a successful program for adults with congenital and valvular heart disease.


The last activity of the day was the annual PICS achievement award. This year the award went to Dr. Philipp Bonhoeffer for his innovative work in congenital heart disease, specifically, for his development of percutaneous pulmonary valve implantation that paved the way for the percutaneous valve replacement/repair era we are witnessing.

Tuesday, September 12th, 2006 was the second full day of the meeting. There were two parallel sessions in two different ballrooms, one for congenital heart disease and the other one for valvular/structural heart disease. To each room, there were live cases transmitted via satellite from around the world. To the adult arena, cases were transmitted from Chicago and Vancouver. For the first time in a US course, a percutaneous aortic valve implantation was transmitted live from St. Paul's Hospital performed by Dr. John Webb, the leader in the world in this technique. This was followed by a case of Mitral valve repair also from Vancouver by Dr. Webb. Again, this case was the first to be transmitted live to a US course. From Chicago, I transmitted to the adult arena two cases: an ASD and a PDA device closure in two patients demonstrating the imaging techniques and the devices used.

To the Pediatric Arena, live cases were transmitted from Toronto Sick Kids with Dr. Benson who performed a percutaneous pulmonary valve implantation; from Columbus, OH where Drs. Cheatham and Galantowicz transmitted a case of hybrid therapy for a neonate with HLHS, while Dr. Amin performed three cases of LPA angioplasty, large ASD closure in a small child under TEE and ICE guidance to demonstrate the two techniques, and a coronary artery fistula that the attendees and the moderators felt that Dr. Amin should wait and not do the case, which he did! I finally transmitted a case of RVOT stent implantation in a child with severe conduit obstruction after a Rastelli operation.

That day, a total of 12 cases were transmitted to both arenas.

In terms of lectures: the adult arena focused Tuesday on the aortic valve where over 12 lectures/debates/discussions addressing the various issues related to the aortic valve. The list of the speakers was very impressive including Drs. Bob Bonow, Michael Mack, Allan Stewart, Ted Feldman, Marty Leon, Jeff Moses, Alain Cribier, George Hanzel, Eberhard Grube, Craig Smith, Donna Buckley and a roundtable discussion that included Marty Leon, Gregg Stone, Peter Block, Larry Wood, Amr Salahieh, Jacque Seguin and Raoul Bonan. Dr. Webb gave his aortic valve talk via satellite from Vancouver.

In the afternoon sessions in the adult arena, the focus was on the PFO/ASD closure. Dr. John Carroll talked about the use of imaging modalities during PFO and ASD closure; this was followed by Bob Sommer who talked about the basics of PFO closure; Jon Tobis talked about strokes and PFO; Jose Suarez de Lezo talked about closure of ASD in adults and Horst Sievert talked about new PFO and ASD closure devices. Of note, on Monday, Horst transmitted live cases from Frankfurt, and traveled the whole day to come and give his talk. It was most certainly, appreciated by all. The last activity of the day in the adult arena was an excellent debate between Gregg Stone and Paul Kramer about PFO closure…I was told that Gregg won that debate calling for randomized trials before we can recommend PFO closure to prevent stroke recurrence.

The pediatric arena also had a very busy schedule of lectures and debates. The list of speakers and topics included: A mini symposium on stents given by Felix Berger, Frank Ing, Chuck Mullins, Evan Zahn and David Bichell. Another mini symposium on ASD and PDA closure was also organized and speakers discussed various issues related to ASD and PDA including: echocardiographic evaluation in children by Charlie Kleinman, the Helex device by Geoff Lane; the Amplatzer device by Jozef Masura; closure of large PDAs by Krishna Kumar and closure of PDAs in premies by Neil Wilson.

Donna Buckley, MD from the FDA gave a talk about the appropriate trial design to seek device approval in the pediatric age


group. This was followed by an interesting debate between Evan Zahn and Emile Bacha on coarctation angioplasty in children… this was certainly a lively debate.

During lunch there were some parallel sessions: the popular "my nightmare case in the cath lab." Needless to say, the moderators of this popular session (Neil Wilson and Mike Mullen) made this the most crowded session. Another session was on fetal intervention that featured the pioneering work of Wayne Tworetzky and Achi Ludomirsky.

Also, in the afternoon, there were parallel sessions for nurses and techs organized by Ms. Sharon Hill. On Tuesday, the focus was on Adult Congenital Heart Disease. In this session, Dr. Chuck Mullins, Dr. Stephen Cook, Ms. Elizabeth Sparks and Mr. Russel D'Sa gave excellent presentations addressing various issues related to the adult with CHD.

The last day, September 13th, was equally busy in both arenas. In the adult arena, live cases were transmitted from Columbia Medical Center in New York City and from Evanston, IL. Dr. Bob Sommer performed three excellent cases and Dr. Ted Feldman also performed 3 excellent cases of valvular and structural heart disease intervention. To the Pediatric Arena, Drs. Tom Jones, Mark Reisman performed three cases from Seattle and Dr. David Balzer performed also three cases from St. Louis. All cases that day were excellent and educational and the attendees enjoyed them tremendously.


From lecture point of view, in the adult arena the focus that day was on the mitral valve and its diseases and the various new technologies available to treat MR/MS.

Drs. Allan Schwartz, Roberto Lang, Marc Gillinov, Francesco Maisano, Steve Bolling, Igor Palacios, Howard Herrmann, Gregg Stone, Maurice Buchbinder, Peter Block, John Webb, George Hanzel, Stephen Ellis, and Allan Stewart gave excellent talks addressing the various aspects of mitral valve disease and therapeutic tools. This was followed by a roundtable discussion featuring Marty Leon, Gregg Stone, Peter Block, Don Bobo, Ferlyn Powell and David Reuter. Discussion focused on the industry and medical community, and what can be done to bring these tools to patient side. A debate between Allan Schwartz and Michael Mack regarding mitral valve therapeutic techniques took place a day before. The debate was excellent and friendly. The last session for the adult arena was focused on the VSDs in adults given by Jo De Giovanni, PDAs in adults by Ziyad Hijazi, LAA imaging and closure by Roberto Lang and David Holmes, coronary fistulas by Mazeni Alwi and paravalvar leaks and their management by Jean Francois Piechaud.

In the pediatric arena, there were many topics discussed including a mini symposium on Hybrid intervention featuring John Cheatham and Mark Galantowicz who discussed their pioneering work in HLHS, David Bichell and Emile Bacha talked about intra operative stent implantation and VSD closure. Then a mini symposium on VSD closure was organized and this featured the echocardiographic evaluation of VSD by Norm Silverman, techniques and results of VSD closure using the Amplatzer device by John Bass, closure of muscular VSD by Basil Thanopoulos, use of Nit Occlud for VSDs by TP Le.

Then, a very interesting and lively debate between Joaquim Miro and Redmond Burke about PmVSD closure, this was indeed a fun debate!

The last two talks of the meeting were given by John Cheatham on cutting balloons and Fontan Completion in the cath lab by Lee Benson.

The parallel sessions that day included a luncheon session on critical aortic valve disease beyond fetal life. This session featured Norm Silverman who discussed the echocardiographic features of this disease, Gil Wernovsky who discussed the ICU management of such patients, Shak Qureshi discussed the catheter option technique and Redmond Burke discussed the surgical management of these children. The other parallel session was focused on "Challenges in the sick neonates". In this symposium moderated by Gil Wernovsky and Sharon Hill, speakers touched on cardiac catheterization and interventions in the critically ill neonate by Evan Zahn; perfusion modifications for cardiac catheterization while on ECMO by Vince Olshove; device closure in the neonate and small infants by Ziyad Hijazi, and premounted stents and hybrid procedures to palliate critically ill neonates by Sharon Hill.

After full three and a half days of intense lectures and live cases, the attendees were invited to the Gala night party that was held Wednesday September 13th, 2006 at the Bellagio Hotel. During the Gala, raffle ticket (30 GB iPOD© courtesy of Congenital Cardiology Today) was given to the person who traveled the longest to come to PICS. Also, AGA Medical and B. Braun held their annual raffle to give four attendees free four registrations for PICS-2007! Then the band which was excellent, entertained the attendees until midnight!!

So, we hope to see you all at PICS-XI to be held again at the Bellagio, in Las Vegas July 22-25th, 2007.

**~CCT~**

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## Ziyad M. Hijazi Receives the PICS Founder Award

*By CCT Staff*

Ziyad M. Hijazi, MD, MPH, Professor of Pediatrics and Medicine at the University of Chicago received the PICS Founder Award at the Pediatric and Adult Interventional Therapies for Congenital and Valvular Heart Disease – PICS 2006, held in the Bellagio Resort in Las Vegas, September 10-13, 2006. Dr. William Hillenbrand, a Course Co-Director with Hijazi since the first PICS, presented the award. PICS, now marking its first decade, has become an annual institution, and is the premier international forum for the advancement and teaching of interventional procedures for congenital heart disease.

The hallmark of PICS is numerous live interactive case demonstrations from multiple international venues. PICS attendance has grown to approximately 1000 international congenital cardiologists and surgeons from its relatively humble beginning in Boston with about 80 American pediatric cardiologists attending the first PICS in 1997.

## MEDICAL NEWS, NEW PRODUCTS AND INFORMATION

### Medtronic Announces CE Mark Approval of the World's First Transcatheter Valve Replacement System For Structural Heart Disease

Signaling a new frontier in the use of transcatheter valve technology, Medtronic, Inc. has announced that it has received CE Mark approval for its Melody™ Transcatheter Pulmonary Valve and Ensemble™ Transcatheter Delivery System. The system is the first of its kind worldwide to treat congenital patients with structural heart disease requiring pulmonary heart valve replacement.

The Medtronic Melody™ Transcatheter Pulmonary Valve and Ensemble™ Transcatheter Delivery System is designed to benefit patients with congenital heart defects involving the connection between their right ventricle and pulmonary artery. Oftentimes, this type of defect requires open heart surgery early in life to implant a prosthetic valved-conduit to establish adequate blood flow from the heart to the lungs. However, the functional life span of these conduits is relatively limited, and as a result most patients with this type of defect are committed to multiple open heart surgeries over their lifetime. The Melody valve and Ensemble system provide a non-surgical means to restore effective valve function and prolong the functional life of prosthetic conduits, thereby reducing the number of open heart surgeries for these patients throughout their lifetime.

"While the population that will benefit from this new technology is relatively small, Medtronic is committed to patient-centered lifetime management of congenital heart disease," said Oern Stuge, MD, senior VP and president of the Cardiac Surgery business at Medtronic. "The Melody Valve and Ensemble System are examples of Medtronic's commitment to evolving the science and focus of structural heart disease. This focus further demonstrates our dedication to delivering innovative valve technology that addresses unmet patient needs. By providing less invasive options for physicians and therefore eliminating even one open heart procedure for a patient, we improve that patient's quality of life immeasurably."

Medtronic has partnered with Professor Philipp Bonhoeffer, professor and chief of cardiology and director of the Catheterization Laboratory, Great Ormond Street Hospital for Children (GOSH) in London, and NuMed, a pediatric catheter company in the development of the Melody Valve and Ensemble System. A pioneer in transcatheter valve technology, Professor Bonhoeffer implanted the world's first transcatheter valve in September 2000. To date, more than 150 patients worldwide have been implanted with the Melody Transcatheter Pulmonary Valve.

Transcatheter valve technology represents a less invasive means to treat heart valve disease and is designed to allow physicians to deliver replacement valves via a catheter through the body's cardiovascular system, thus eliminating the need to open the chest. Traditionally, open heart surgery has been required to correct the problem and it is not unusual for a patient to undergo multiple, open-heart surgeries during their lifetime. Patients with this condition tire very easily, as the heart over-exerts itself trying to get oxygenated blood throughout the body.

"To reopen the chest many times is obviously uncomfortable for the patient but also comes with risk. By using the Melody Valve and Ensemble System, we can now avoid the open chest procedure and instead perform the same procedure safely, quickly, and more comfortably," said Prof. Bonhoeffer.

CE Mark Approval gives Medtronic the ability to train and educate physicians in Europe in preparation for implanting the Melody™ and Ensemble™ System so that more patients can benefit from this innovative treatment. With proper training in place, Medtronic will work with hospitals across Europe in gaining access to this new technology.

While the Melody Valve and Ensemble System are not currently available in the United States, Medtronic is working with the U.S. Food and Drug Administration to pursue plans to apply this new approach in transcatheter valve therapy for patients in the U.S.

For more information about the product and treatment visit www.melody-tpv.com.

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### The International Children's Heart Foundation Needs Volunteer Pediatric Cardiac Specialists for Upcoming Missions

William M. Novick, MD of the University of Tennessee Health Sciences Center, and Founder and Medical Director of the International Children's Heart Foundation is looking for volunteers for the following missions:

- Armed Forces Institute of Cardiology/ National Institute of Heart Disease, in Rawalpindi, Pakistan; Dec. 2-16, 2006: Pediatric Cardiac Anesthesiologist (1) and pediatric cardiac perfusionist (1).
- Beijing Children's Hospital, Beijing, China; Jan. 6-20, 2007: Pediatric Cardiac Anesthesiologist (1).
- Armed Forces Institute of Cardiology/ National Institute of Heart Disease, in Rawalpindi, Pakistan; Jan. 27- Feb. 10, 2007: Pediatric Cardiac Anesthesiologist (1), Pediatric Intensivist (2), Pediatric Cardiologist (1).

Interested individuals should contact Martina Pavanic, Volunteer Coordinator, at martapavanic@aol.com.

# BERRY SYNDROME IN A NEONATE: REPORT OF A CASE SUCCESSFULLY REPAIRED IN A ONE-STAGE OPERATION

*By Marcì Marcello, MD; Calvaruso Davide, MD; Cipriani Adriano, MD; Ocello Salvatore, MD; Salviato Nicoletta, MD; and Marcelletti F. Carlo, MD*

**Case Report**

A severely ill infant, weighting 3,1 Kg, was referred to our Institution soon after the birth for severe cyanosis, metabolic acidosis (pH = 7,1) and anuria (creatininemia =3,5 mg/dL). Physical examination revealed tachypnea and a systolic murmur while femoral pulses were weak. Radial and femoral arterial pressures, invasively monitored showed a difference of about 30 mmHg. Similarly oxygen saturation was higher in upper limbs. Echocardiography showed interruption of the aortic arch, type A, according to the classification of Celoria and Patton. Thus flow in the descending aorta was provided by a patent ductus arteriosus. Furthermore, aortopulmonary window and anomalous origin of the right pulmonary artery from ascending aorta were associated.

Correction of acidosis, PGE1 and dopamine infusion and diuretics allowed rapid recovery with stabilization of hemodynamic conditions and balance of oxygen saturation and arterial pressure in upper and lower limbs, with subsequent recovery of renal function.

Cardiac catheterization and angiocardiography confirmed the presence of aortic arch interruption associated with PDA, aorto-pulmonary septal defect and anomalous origin of RPA.

At 5 days of age the baby underwent total repair with deep hypothermic circulatory arrest a 14°C nasopharingeal.

After aortic cross-clamping distal to the defect, cardioplegia was infused through an arterial cannula into the ascending aorta that was incised transversely showing a very large aorto-pulmonary window. The coronary artery origins were normal, the pulmonary and aortic valve leaflets were identified. The right pulmonary artery originated directly from the right-lateral wall of the ascending aorta. After the great vessels were divided, both aortic and pulmonary defects were closed using two different pericardial patches with continuous 6/0 polypropylene suture. The PDA was divided and all ductal tissue was removed. Right pulmonary artery was excised from aorta and then anastomosed to the main pulmonary artery. The mobilized descending aorta was end-to-end anastomosed to aortic arch with an 8 mm goretex prosthetic tube. Wean from cardiopulmonary bypass was early obtained.

The recovery from operation was uneventful and the patient was discharged home on the 20th postoperative day.

**Comment**

Aorto-pulmonary septal defect is a rare malformation: its incidence ranges from 0.2% to 0.6 % in patients with congenital heart diseases, according to Kutsche and Van Mierop.

It may be associated in about 25% of patients with interruption of aortic arch, usually type A of Celoria and Patton classification .

Etiological factors involved in the development of aortic arch interruption (AAI) and aorto-pulmonary defect (APW) are not well defined. Though both abnormalities involve the aorta, they have different embryological pathogenesis. APW should be caused by lack of development of aorto-pulmonary septum in the aortic sac, which appears late in embryogenesis and is believed to be caused by reduction of blood flow toward the descending aorta during intrauterine life because of the frequent association with intra-cardiac lesions with obstruction to the left ventricular outflow tract and/or VSD. This hypothesis has been confirmed by experimental observations in animal embryos, in which aortic interruption was caused by restriction of the conotruncus. According to this hypothesis AAI should be the consequence of reduction of blood flow into the ascending aorta. In addition, a genetic disorder could play a role in development of interruption of aortic arch, as demonstrated by the association of this abnormality with Di George syndrome. Whereas, aorto-pulmonary septal defect has never been reported in patients with chromosomal delection in 22q11. In fact, development of aortic arch and of aorto-pulmonary outflow tract are influenced by migration of neural crest cells to the branchial arches and to the aortic sac. Removal of the cardiac neural crest region in the animal embryo can determine truncus arteriosus and type A interruption, but not defect of aorto-pulmonary septum.

Several classifications of APW have been proposed according to the size and location of the defect. In Richardson et al's classification three types of aorto-pulmonary defect are described: type I, the most common, is a defect localized between ascending aorta and pulmonary trunk, in the type II the defect extends distally toward the bifurcation of the pulmonary artery, the type III is an anomalous

origin of the RPA or hemitruncus arteriosus. According to Richardson's classification, Berry syndrome would be a combination of two different types of APW. Actually, hemitruncus is not considered a form of aorto-pulmonary defect by other authors. Mori et al. for example, distinguish three forms of APW: "proximal defect" is just above the semilunar valves, "distal defect" is near the bifurcation of pulmonary artery, and "total defect" involves most of the ascending aorta and of the main pulmonary artery.

In the majority of cases, aortopulmonary septal defect is frequently associated to other lesions, such as type A interruption of aortic arch, anomalous origin of right coronary artery from pulmonary artery, tetralogy of Fallot, transposition of great arteries, right aortic arch and anomalous origin of the RPA. Moreover association with ventricular septal defect, d-transposition, pulmonary or aortic atresia have also been described. Patients affected by APW and associated lesions have a worse prognosis. The association of aortopulmonary septal defect, PDA and anomalous origin of RPA from ascending aorta is extremely rare and was reported in 1982 by Berry et al. This potentially lethal lesion may be a tough diagnosis for the cardiologist, because an accurate echocardiographic identification is not always obvious. Often a cardiac catheterization is needed to confirm the diagnosis. An early surgical treatment is recommended to avoid the development of irreversible obstructive pulmonary vascular disease.

Boonstra et al. reported the first surgical repair of such syndrome in 1992. To our knowledge, since then, only a few other cases of Berry syndrome, surgically treated, have been reported in literature.

**Summary**

We report a case of Berry syndrome, a rare combination of aorto-pulmonary malformations, in a neonate successfully treated with primary surgical repair.

See authors for reference information.

**~CCT~**

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**CONGENITAL CARDIOLOGY TODAY**


(ISSN 1554-7787-print; ISSN 1554-0499-online)
*Published monthly. All rights reserved*

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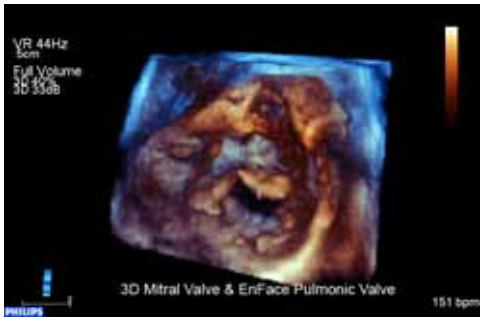


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