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Trans-catheter Closure of Patent Ductus Arteriosus (PDA) in Extremely Low Birth Weight (ELBW) Premature Infants: A New Treatment Option for an At-Risk Population

By Evan M. Zahn, MD; Ruchira Garg, MD; Alistair Phillips, MD

Introduction

Patent Ductus Arteriosus (PDA) is the most common cardiovascular anomaly in the neonatal period with an incidence as high as 80% in Extremely Low Birth Weight (ELBW) infants defined as a birth weight <1200 gm. The presence of a hemodynamically significant PDA in this population is associated with significantly increased morbidity including necrotizing enterocolitis, chronic lung disease and intraventricular hemorrhage.[1-7] A recent report suggests that mortality in neonates born at <29 weeks gestation may be increased by more than eight times in those with a PDA versus those without,[7] and it is estimated that 60-70% of infants born <28 weeks gestation will undergo some form of therapy for PDA. Despite these staggering numbers, definitive management algorithms for this lesion remain controversial, largely due to the fact that both traditional medical and surgical therapies for PDA in this population have been associated with significant adverse events.[8-14] Cox (cyclooxygenase) inhibitors such as indomethicin have been associated with permanent or transient alterations in renal function, necrotizing enterocolitis, gastrointestinal perforation and impairment of cerebral blood flow velocity.[8-10] Additionally, these medications are thought to be most effective in infants with gestational ages between 32-36 weeks, with failure rates quoted as high as 43% in ELBW infants weighing < 800 gm.

Surgical ligation, while successful in nearly all cases involves a limited left thoracotomy and is typically reserved for patients who fail medical therapy. Ligation has been associated with significant procedural complications including: pneumothorax, hypothermia, bleeding, phrenic nerve palsy, wound infection, vocal cord paralysis and thoracic scoliosis.[11-14] While it is generally accepted that surgical PDA ligation is more effective than medical therapy, to date, it has not been shown to positively impact survival and in fact, may result in significant long-term sequelae.[15-16] Several recent large studies have suggested that ligation is independently associated with the development of chronic lung disease and neurosensory impairment.[15,16] Undoubtedly, this is a complex high-risk patient population, often with multi-organ system disease of which PDA may have varying degrees of importance; however, one can not help but speculate that if a more predictable, less invasive therapy, associated with fewer adverse events could be developed, the ELBW neonatal population would benefit.

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Transcatheter PDA closure was first described nearly 4 decades ago by Porstmann[17] and has become the procedure of choice for infants >5 kg, children and adults around the world. The safety and efficacy of this therapy has been demonstrated in numerous studies and several devices have been successfully developed and approved specifically for this use around the world including in the United States. Importantly, a large number of devices designed primarily for other purposes have also been used successfully to close PDA of varying sizes and shapes including, coils, septal occluders and vascular plugs. A recent report from the Congenital Cardiac Catheterization Outcomes Project (C3PO) examining 496 transcatheter closure cases of PDA from 8 centers using PDA plugs or coils, reported successful closure rates of 97-99% with a severe adverse event rate of 2.2% and 0% mortality.[18] Of note, however, infants <6 kg and <6 months of age had significantly higher significant adverse event rates (10%) with longer procedure times and contrast dose emphasizing the technical difficulty of this procedure using standard techniques and devices in smaller patients. The findings of this multi-center publication epitomize the fact that suitable devices and catheter techniques have, until very recently, prevented this therapy from being widely applied to ELBW infants. But landmark inroads are being made.

In 2010 Francis and colleagues reported on 8 preterm infants (median weight 1,110 gm) who underwent successful catheter-based PDA closure using multiple steel coils delivered with bioptome assistance.[19] These procedures were performed in the catheterization laboratory where fluoroscopy and aortic angiography were used to guide the closure procedure similar to the standard technique used in older children and adults. The authors noted that while they had a high success rate, specific ductal anatomy was required for success, namely a large aortic ductal ampulla which they estimated, was only present in approximately 10% of their population (making 90% of patients unsuitable for this approach). Nonetheless, this landmark study demonstrated that transcatheter ductal closure could be adapted to this in-need, at-risk population. Shortly thereafter, Bentham et al described a novel technique of bedside neonatal PDA closure using echocardiographic guidance to implant either detachable coils or the Amplatzer Duct Occluder II (ADO II).[21] While this approach was appealing in that it avoided transport out of the Neonatal Intensive Care Unit (NICU), exposure to ionizing radiation and contrast injection, there were some notable drawbacks. These included the need for femoral arterial vascular access and lack of fluoroscopy should device repositioning or retrieval be needed. Based upon this report, we embarked on our first case of catheter-based PDA closure in a 2400 gm neonate using an identical approach and, like the authors, we were struck by the advantages of performing the procedure bedside, ease of implantation from a retrograde arterial approach, good visibility of the device, in the case an Amplatzer Vascular Plug II (AVP II - the ADO II was not available in the United States at that time) and satisfying result. However, like the authors of the original article, we too, were concerned about placing a 4 Fr catheter into the femoral artery of a small neonate and felt that this approach would not be feasible in ELBW neonates due concerns over limb ischemia. Additionally, we felt that the complete absence of fluoroscopy would limit our ability to manage device complications such as malposition or embolization. After discussion among our multidisciplinary team (cardiology, non-invasive cardiac imaging, cardiac surgery, neonatology and intervention) we developed the technique described in this report, which uses a transvenous approach to implant the commercially available AVP II (St. Jude Medical, Minneapolis, MN) into the PDA's of ELBW neonates with immediate conversion to standard surgical ligation should that be necessary.

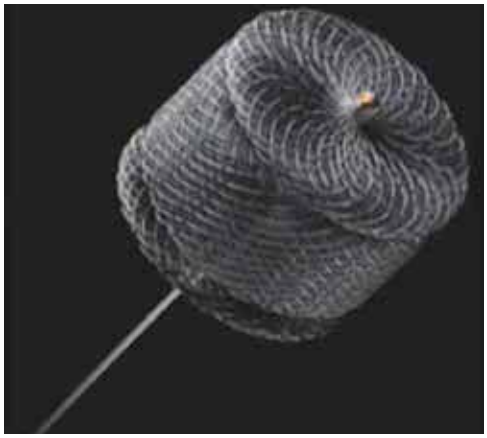

Figure 1. Commercially available Amplatzer Vascular Plug II (St. Jude Medical, Minneapolis, MN). The small diameters and lengths of this device, easy deliverability and excellent fluoroscopic and echocardiographic features of this device are attributes desirable in a PDA device in small neonates.

The Device

The AVP II (Figure 1) is a densely woven, self-expanding nitinol plug composed of 2 layers of 144 braided nitinol wires constructed as 2 outer disks and a central plug of equal diameter. Some advantages of this device as they pertain to this particular procedure include: excellent fluoroscopic and echocardiographic visibility, ability to reposition/remove multiple times as needed, relatively short length (3-6 mm diameter devices have an unconstrained length of 6 mm), deliverable through a 4 F sheath, relatively thin and soft delivery cable and rapid vascular occlusion. The manufacturer recommends choosing a plug with a diameter at least 20% larger than the target vessel. Importantly for the current application, the final length of the deployed device is dependent on the degree of oversizing, i.e. the more over-sized the device (more constrained) the greater the implanted length will be.

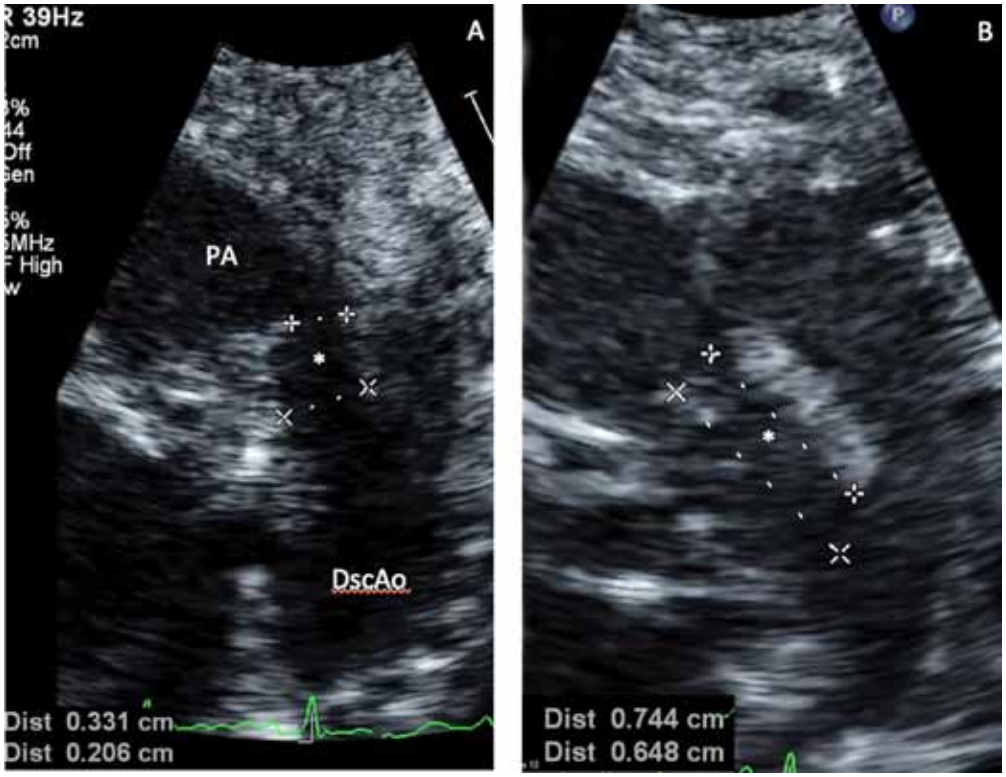

Figure 2. Two-dimensional transthoracic images of the pertinent anatomy obtained form the high left parasternal position. The PDA (*) diameters (A) and length (B) from the pulmonary artery (PA) to descending aorta (DscAo) are measured to determine candidacy for transcatheter device closure. With a minimum PDA length > 6 mm and narrowest diameter of 2 mm, this patient was selected for device closure and a 3 mm AVPII was successfully implanted.

Our Current Approach to ELBW Neonatal PDA Closure

All cases referred by neonatologists for PDA closure undergo extensive quantitative echocardiographic evaluation to assess suitability for transcatheter closure (Figure 2). This involves quantitative measurements of ductal diameter and length made in a variety of parasternal views. If ductal length is ≥6 mm and minimal diameter ≤3.5 mm, patients are considered suitable candidates for an attempt at transcatheter closure. Those with unsuitable anatomy undergo standard surgical ligation at the bedside. To date, age, weight and degree of illness have been not been used to exclude or select patients for this procedure. After we obtain informed consent for both catheter and surgical closure (surgery is scheduled as a fallback under the same anesthetic should catheter closure not be possible, deemed sub-optimal or if a complication arises), our patients are transported to the catheterization suite. All cases are performed under general anesthesia with endotracheal intubation using the lowest possible fluoroscopy settings. A 4F introducer sheath (Daig, St. Jude Medical, Minneapolis, MN) is placed in the right femoral vein using a 21-gauge butterfly needle, floppy tipped 0.018 guide wire (Micropuncture set, Cook Inc, Bloomington, IN) and standard Seldinger technique. Intravenous heparin (100 mg/kg) and prophylactic antibiotics (Cefazolin, 20 mg/kg) are administered in all cases (unless contraindicated) and activated clotting time monitored and maintained between 200-250s. Under fluoroscopic guidance a 4F balloon wedge catheter (Arrow International, Reading, PA) is advanced to the mid-right atrium, the balloon is inflated and the stiff end of a 0.018 standard guide wire (Cordis, Miami Lakes, FL), which is shaped into a tight curve is advanced to the tip so as to direct the catheter towards the tricuspid valve (Figure 3, Video 1-Deployment). This minimizes catheter manipulation in the right atrium and allows for atraumatic passage of the catheter into the right ventricle where it can either be passed into the main pulmonary artery or a soft guide wire passed through it into the pulmonary artery and down the PDA. After passing a 0.014 floppy-tipped coronary guide wire (Hi-torque ALL STAR, Abbott Vascular, Santa Clara, CA) across the PDA, down the descending aorta and into a femoral artery, the balloon end-hole catheter and introducer sheath are removed and replaced with a 4F long hydrophilic sheath (Flexor Check Flo Introducer 4 F x 45 cm, Cook Inc., Bloomington, IN), which is advanced through the right heart, across the PDA into the descending aorta (Figure 4). Gentle external pressure over the femoral artery where the tip of the coronary guide wire is located can be used to stabilize the guide wire, and provide the stability of a veno-arterial wire rail system (without exteriorizing the wire) for passage of the long sheath. After removing the dilator and guide wire from the delivery sheath, transthoracic echocardiographic images of the ductal region and appropriate anatomy including the para-ductal descending aorta and origin of the left pulmonary artery (LPA) are obtained and used to guide device placement. The AVP II is prepared in the standard fashion and advanced to the tip of the delivery sheath under fluoroscopic guidance. The sheath is brought back to the aortic end of the PDA before deploying the distal disk. Efforts are made to deploy the aortic disk directly into the aortic ampulla of the ductus. Slow controlled traction on the sheath results in deployment of the remaining central portion and pulmonary end of the device into the PDA. Again, efforts are made to deploy as much of the device into the actual ductus and leave as little in the main pulmonary artery as possible. A detailed echocardiographic assessment including color and spectral analysis of flow in the PDA, LPA and descending aorta are made prior to releasing the device from the delivery cable. When concern arises regarding obstruction of flow into the LPA or descending aorta, the device is repositioned using either gentle traction (aortic obstruction) or partial recapture of the proximal device with redeployment in a slightly more distal location (LPA obstruction). When the decision is made that the device has closed the PDA completely and there is no impingement on LPA or aortic flow, the device is released from the delivery cable and repeat echocardiographic assessment is made.

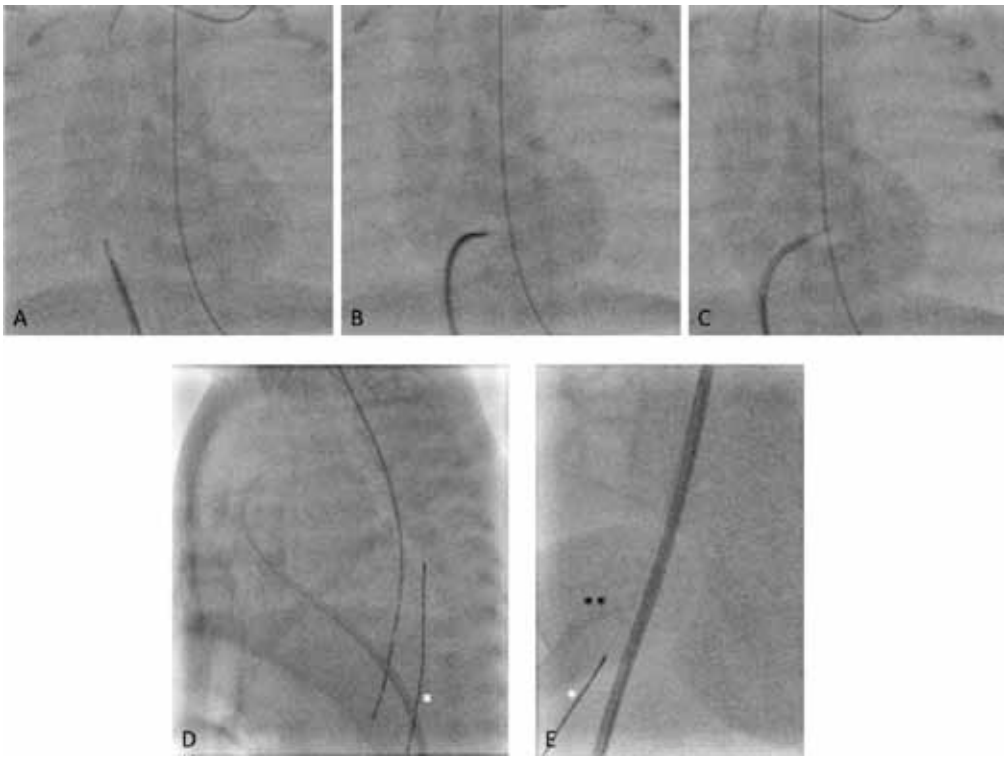

*Figure 3. Minimizing catheter manipulation in an ELBW infant. (A) A 4F balloon tipped catheter is placed in the mid-right atrium. (B) The pre-shaped end of a stiff 0.018" guide wire is advanced to the tip of the catheter curving the balloon tip toward the tricuspid valve. (C) Once the catheter has passed into the right ventricle, the stiff wire is removed and (D) replaced with a floppy tipped coronary guide wire (white *) which is advanced on the lateral projection out the right ventricular outflow tract, across the PDA and down the descending aorta. (E) The guide wire tip (white *) is advanced into the right femoral artery where gentle manual pressure (black **) can be placed to stabilize the wire and create an arterio-venous loop.*


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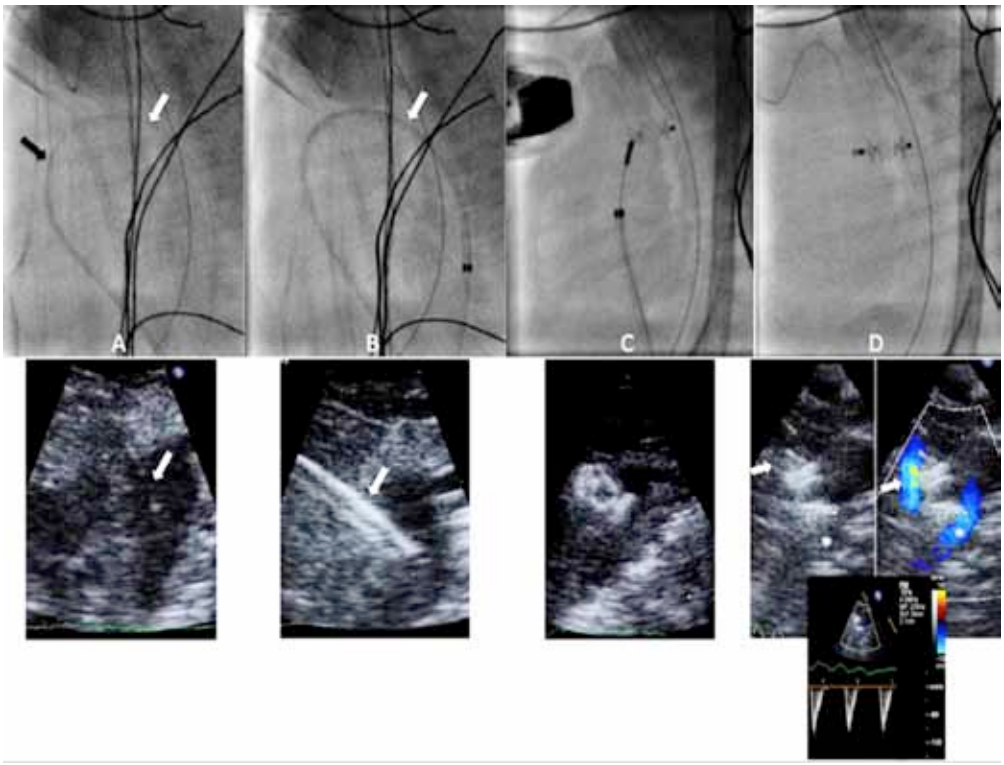

Figure 4. Device implantation sequence. Lateral fluoroscopic (top row) and corresponding transthoracic echocardiographic images (bottom row) of the implant procedure. (A) A 0.014 guide wire (white arrow) has been advanced from the femoral vein across the PDA into the descending aorta and a 4 F long sheath (black arrow) is being advanced over the guide wire through the right heart. (B) Once the dilator has been removed, the delivery sheath (white arrow) can be seen clearly by both imaging modalities as it crosses the PDA. (C) Following deployment, but prior to release from the delivery cable, all 3 components of the AVPII can be visualized and a careful assessment of position made. (D) Both 2 dimensional, color and pulsed wave Doppler are utilized to ensure that the PDA is closed and that normal flow is maintained in the LPA (white arrow) and descending aorta (*).

At case completion, the sheath is removed, hemostasis obtained with manual compression and the babies transferred back to the NICU to receive 2 further doses of intravenous antibiotics.

Follow-up

Follow-up echocardiography and chest radiographs are obtained within 24 hours of the procedure and as clinically indicated thereafter. Particular attention is paid to device position and the possible development of left pulmonary artery or descending aorta obstruction.

Results

At the time of this writing, we have attempted this procedure in 9 premature neonates whose demographic data are shown in Table 1. Median gestational age and weight at birth were 27 weeks (26-33 weeks) and 900 grams (440-2480 gm), respectively. Median age and weight at the time of the procedure were 22 days (16-80 days) and 1220 grams (870-2240 gm), respectively. All patients had failed at least one course of COX inhibitors (or were not considered candidates for medical therapy) and were being considered for surgical ligation. Device closure was successful in 8/9 infants with procedural details shown in Table 2.

There was no correlation between patient size and fluoroscopy or procedural times. In fact, two of the shortest cases involved two of the smallest babies (Patients 3 and 5). Three device related incidents occurred in 3 separate cases, resulting in increased procedural and fluoroscopy times in those infants. Patient 2 who had a large PDA, initially, received a 6 mm AVP which achieved complete ductal closure, but resulted in flow obstruction in both the aorta and LPA, presumably because the constrained length of this device was too long (i.e. it was oversized). The device was recaptured prior to release, removed uneventfully and replaced with a 4 mm device, resulting in complete ductal closure and no obstruction to LPA or aortic blood flow.

Patient 4 experienced device malposition following release from the delivery cable seen in this cohort. Prior to release, the device appeared in excellent position with complete ductal closure and no obstruction to flow in either the LPA or aorta. When the device was released, there was a slight, but obvious movement of the device posteriorly on fluoroscopy (Figure 5). This correlated to an

Table 1. Demographic Data

Patient	Birth Weight (gm)	Gestational age (weeks+days)	Sex	Procedure Weight (gm)	Procedure Age (days)	Ventilator Dependence	Inotrope Dependence
1	2480	31+6	Female	2240	18	yes	yes
2	440	26+4	Male	1610	80	yes	no
3	1050	26+3	Male	1140	16	yes	no
4	1077	26+3	Male	1220	20	yes	no
5	856	26+3	Female	960	21	no	no
6	675	29+0	Female	870	30	yes	no
7	900	28+3	Male	1155	22	no	no
8	610	27+0	Female	1655	78	yes	no
9	1000	26+2	Female	1400	30	no	no

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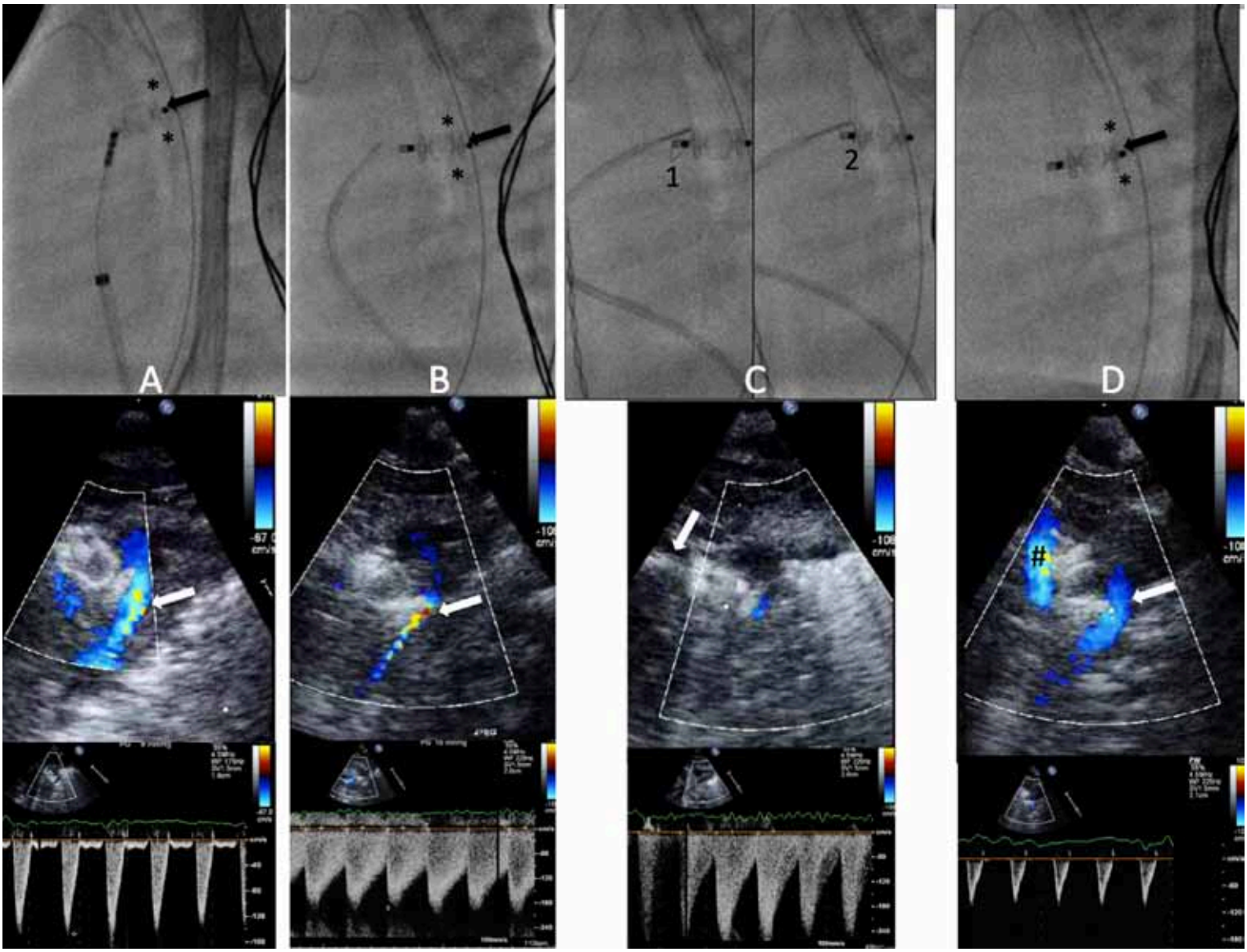
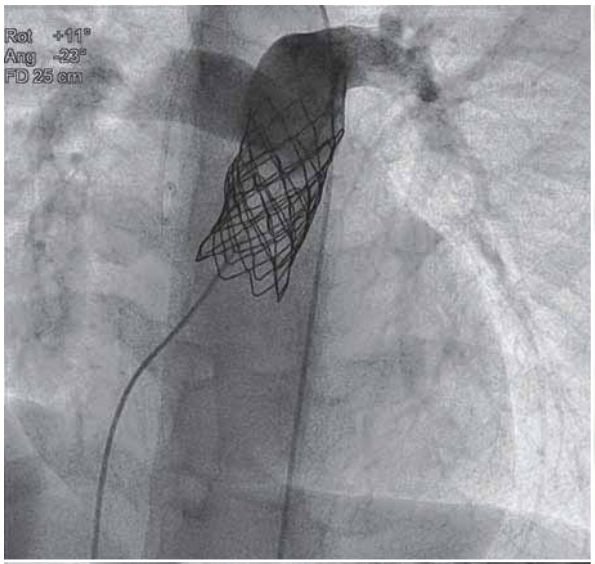
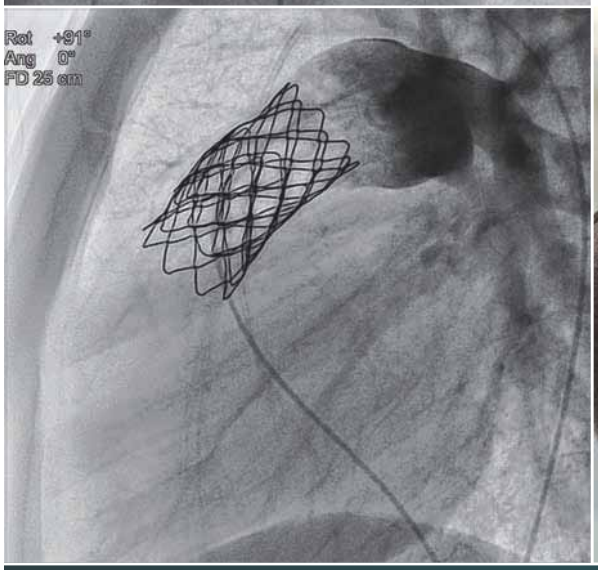


Figure 5. Lateral fluoroscopic (top row) and corresponding transthoracic echocardiographic images (middle row) and Doppler spectral analyses of the descending aorta (bottom row) during PDA closure of a 1220 gm infant with a 4 mm AVP II. (A) Prior to release from the delivery cable the device is in a desirable location with the posterior disk (black arrow) parallel to the posterior margin of the trachea (*) on fluoroscopy and complete ductal closure with normal descending aortic flow by color Doppler (white arrow) and spectral analysis. (B) Following release from the delivery cable, the device (black arrow) has moved 1-2 mm posteriorly in relation to the trachea (*) which results in significant aortic obstruction evidenced by both color Doppler (white) imaging and a marked alteration in the spectral velocity profile. (C) A 5 mm goose-neck snare has been advanced over the device micro-screw (1) in the main pulmonary artery and tightened over the screw (2). Echocardiography clearly shows the snare (white arrow) and an obvious change in device orientation, however, at this point there is still important obstruction to aortic blood flow as demonstrated by the Doppler velocity profile. (D) After applying gentle proximal traction on the device with the snare under continuous echocardiographic monitoring, the device has been returned to its original position as shown fluoroscopically by the parallel relationship of the posterior disk (black arrow) to the posterior margin of the trachea (*). Color Doppler echocardiography demonstrates normal flow in the descending aorta (white arrow) and LPA (#) confirmed by a normal Doppler velocity profile in the descending aorta.

Table 2. Procedural Details

Patient	Procedure Location	Vascular Access	Sheath Size	Procedure Time (min)	Fluoroscopy Time (min)	Minimal Ductal Diameter (mm)	AVP II Size	Successful Transcatheter Closure
1	NICU	Artery	5fr	40	0	1.4	4	Yes
2	Cath lab	Vein	4fr	87	19.5	1.8	4	Yes
3	Cath lab	Vein	4fr	35	7.2	2	3	Yes
4	Cath lab	Vein	4fr	71	18.7	2.7	4	Yes
5	Cath lab	Vein	4fr	33	6	1.2	3	Yes
6	Cath lab	Vein	4fr	51	11.6	3.1	4	Yes
7	Cath lab	Vein	4fr	35	6.1	1.6	3	Yes
8	Cath lab	Vein/Artery	5fr/4fr	147	28.4	3.4	6	Yes
9	Cath lab	Vein	4fr	20	6.1	3.0	4-removed	No





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immediate change in the descending aortic Doppler flow pattern and the 2D echocardiographic appearance of the juxta-ductal aorta consistent with descending aortic obstruction. The patient remained stable and the 4F long sheath was replaced with a 5F long sheath through which a 4mm goose neck snare and catheter were passed. The microscrew of the AVPII was snared easily but it was clear that the device could not be recaptured using this system. Steady gentle retraction on the snare catheter and sheath were used to slowly manipulate the device into a more proximal location under echocardiographic guidance. When the aortic Doppler flow pattern returned to normal, the micro-screw was released from the snare. The final result demonstrated no obstruction to LPA or aortic flow with complete ductal closure. Patient 8, a 1655 gm infant with the largest PDA in the series (3.4 mm at the narrowest diameter), failed closure with a 4 mm device (excessive leak and device instability prior to release from the delivery system), and thus, a 6 mm AVP II was placed. Upon release from the delivery system, we noticed a similar posterior movement of the device as Patient 4, however, in this case gentle traction with a snare catheter advanced from the venous system alone was not enough to reposition the device. We therefore, placed a second snare from the arterial side (via a 4fr sheath in the femoral artery) on to the distal disk, and by exerting gentle tension upon both ends of the device, we were able to slightly collapse the device and move it into proper position, thereby eliminating any aortic obstruction. The final result was excellent with no arterial complication, however, the procedural and flouroscopy times were considerably longer than other patients in the series.

In the one case where device closure was not possible (Patient 9), a 4mm AVP II was successfully placed and the PDA completely occluded, however it was felt that the LPA was slightly compressed by the expanded device and the risk for the development of LPA stenosis was significant. It did not appear echocardiographically that the device was placed too proximally in the main pulmonary artery resulting in ostial obstruction but rather that the relationship between PDA and LPA was such that the body of the device within the PDA resulted in slight but potentially important long segment LPA compression. The device was therefore removed prior to release from the delivery cable, and the patient underwent uneventful surgical ligation using the same anesthetic.

During this same time period, 3 ELBW neonates were sent for surgical PDA ligation based upon their anatomic unsuitability for the transcatheter procedure.

Discussion

There is little debate that the presence of a PDA in premature infants is associated with important comorbidities and increased mortality. Both medical and surgical therapies for PDA in this population have been associated with significant adverse events[8-14] contributing to a change in practice designed to forestall or forgo definitive treatment of this lesion in this population. Recent advances in catheter and device technology and the pioneering work of several groups have demonstrated that in fact, a catheter-based approach to this lesion is possible even in neonates <1000 gm. The advantages of such an approach are obvious, and include potential avoidance of medications, which are only partially successful, and are associated with significant side-effects, and avoidance of a left thoracotomy and the well-described complications associated of surgical ligation in fragile ELBW neonates.

The AVP II is a relatively new device (released in the United States in 2007), which has been shown to be useful in a wide variety of settings. The particular design feature which facilitates use of this device for ductal closure in premature neonates is that the retention disks on either end of the device are the same diameter as the central occlusion portion. This is in contrast to other occlusion devices such as the ADO II where the retention disks are 6 mm larger than the central occlusive portion. This feature is advantageous in these premature neonates where the terminal ends of any device may interfere with aortic and branch pulmonary artery blood flow. The trade-off of this design feature is a theoretically higher risk of embolization due to smaller retention disks securing the device. While in this initial series we observed no instances of device embolization, with this device configuration and only oversizing the device 1-2 mm greater than the PDA, the potential risk of embolization will need to be carefully scrutinized in a larger series. Additionally, the 2 cases in this series of device malposition after release from the delivery system, highlight the need for improved devices designed specifically for this PDA in this population. The patients treated in this series all had tubular ductal anatomy (Krichenko

Class C or E), an important consideration when evaluating a neonate for transcatheter closure with the AVP II.[21] It has been our personal experience that tubular ductal morphology appears to be more common in ELBW infants than in older children suggesting this device may be appropriate for a larger percentage of premature neonates than coils. In this small pilot series, approximately 70% of premature neonates presenting to our group for PDA closure could successfully undergo this procedure. Using a technique which combines fluoroscopy (primarily for catheter and wire passage) and echocardiography (primarily to guide device deployment and assess result) we have been able utilize a completely transvenous approach in ELBW infants as small as 870 gm while avoiding lengthy exposure to ionizing radiation and contrast injections. The addition of limited fluoroscopy also allows for device repositioning, and potentially removal, after release for the delivery system. The current drawback to this approach is the need to transport these fragile infants to a catheterization suite to undergo this procedure. We are working with industry partners to resolve this important issue by developing a bedside method utilizing a unique bed (Neoforce, Ivyland, PA) and imaging chain which will allow this procedure to be performed bedside in the NICU (Figure 6).

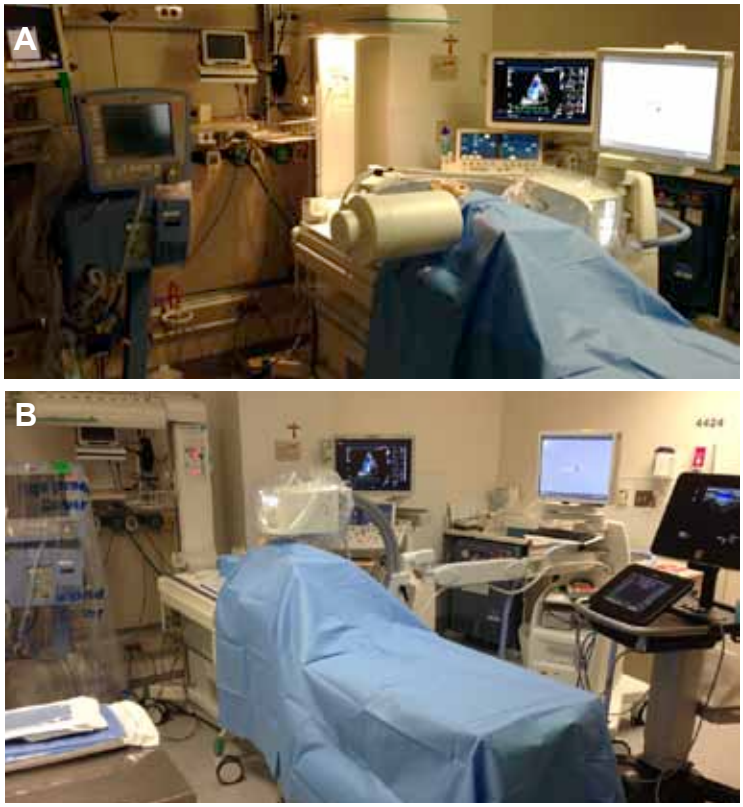

Figure 6 A / B. The Rainbow Flex bed (Neoforce, Ivyland, PA), an open neonatal surgical/procedural bed shown will facilitate the use of a small portable digital C-arm in the neonatal intensive care unit. In the near future this will allow for transcatheter PDA closure and/or conversion to surgical ligation at the bedside, obviating the need to move these fragile patients to the catheterization suite.

While we have described a novel technique to accomplish successful PDA closure in a small series of premature neonates, it is important to recognize that several things could improve this procedure. First and foremost, while the AVP II has worked well in this setting to date, we feel that a shorter and softer device with a softer delivery cable (such as the ADO II AS, St. Jude Medical, Minneapolis, MN, currently available only outside the United States) will improve the safety profile of this procedure and increase applicability to an even larger group of in-need ELBW neonates. This device features not only shorter lengths (2-6 mm), but retention disks slightly larger than the central occlusive portion (1-1.5 mm larger), which would theoretically minimize malpostion or embolization. Several authors have very recently reported encouraging early results using this device in neonates.[22, 23] Going forward, research efforts should be focused on smaller and softer delivery systems (<3F), softer devices with robust seating characteristics, which are easily retrievable.

In conclusion, we report our encouraging early experience using the commercially available AVP II and a novel transvenous technique for PDA closure in premature neonates combining echocardiographic and

fluoroscopic imaging. Further device design and development, as well as modifications of the technique, may ultimately make this a routine bedside procedure performed in the NICU, and change our approach to the ELBW infant with PDA.

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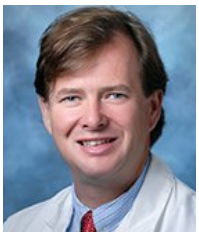

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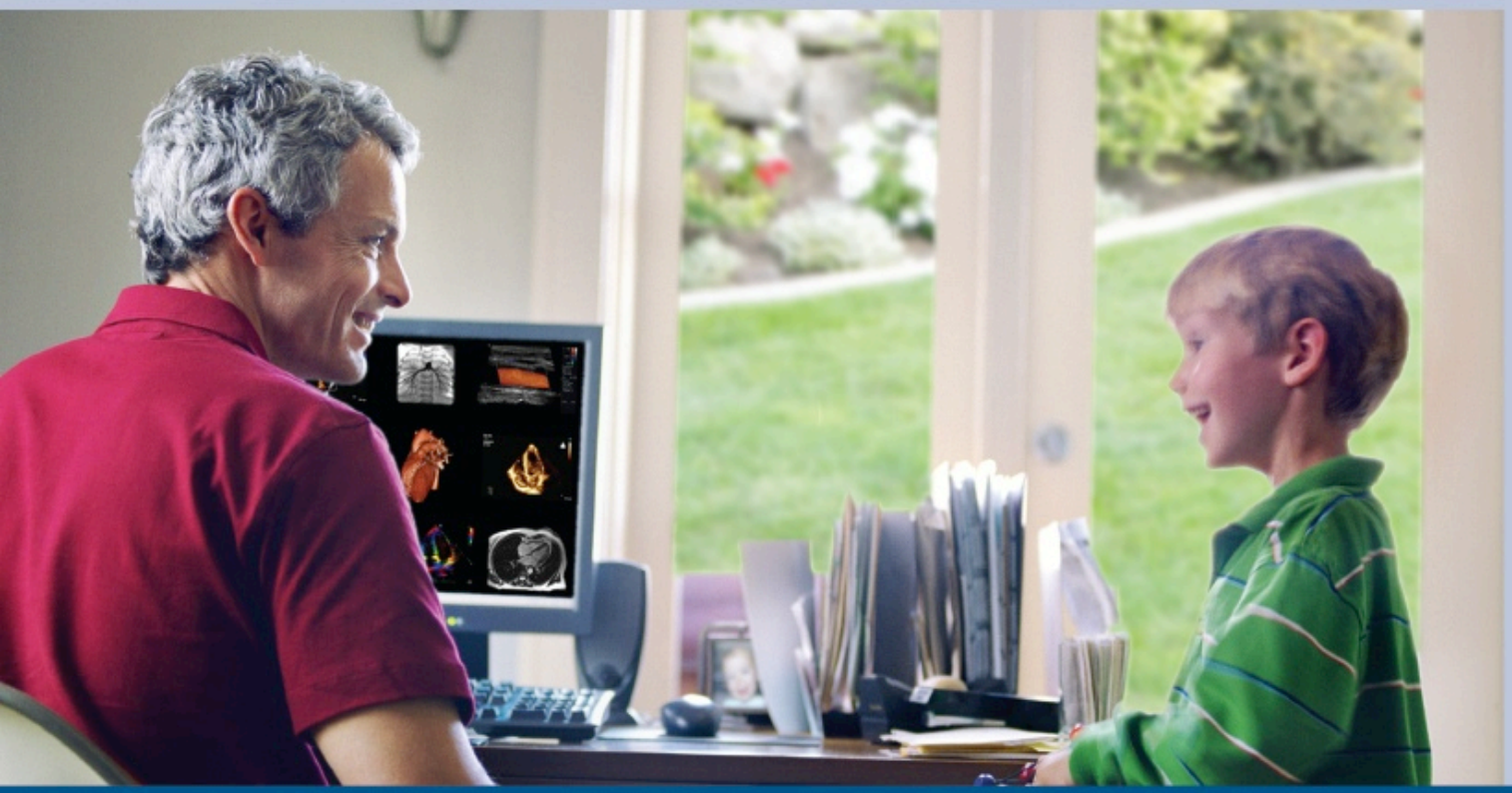

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Arno R. Hohn, MD, Professor of Pediatrics, Children's Hospital Los Angeles University of Southern California 1931-2014

By, Anjan S. Batra, MD; Michael J. Silka, MD

Dr. Arno Hohn

When Arno Hohn passed away in late March 2014, Children's Hospital Los Angeles (CHLA) and the field of pediatric cardiology lost one of its most exemplary leaders and gifted teachers. Arno received his undergraduate degree from Rutgers University and his MD degree from New York Medical College in 1956. His pediatric residency was completed at Children's Hospital of Philadelphia followed by fellowship in pediatric cardiology at Buffalo Children's Hospital. Following military service, he became a faculty member at Buffalo Children's Hospital and then at the Medical University of South Carolina (MUSC). At MUSC he progressed to Professor of Pediatrics and Director of Pediatric Cardiology. In addition, he was interim Director of the Department of Pediatrics at MUSC between 1976 and 1978.

Arno was recruited as chief of the Division of Cardiology at CHLA in 1984, where he served in this role through 1999. During this time, he played a critical role in the development and emergence of the Heart Institute at CHLA as the leading program for cardiovascular care of children in the Western United States. During his tenure, Arno was responsible for the recruitment of a cadre of outstanding physicians, the development of a stellar cardiology fellowship program, and advancement of the care of children with heart disease. His research interests focused on hypertension in pediatrics as well as heart problems in muscular dystrophy, HIV and premature infants.

Among the skills Arno best embraced was the art of the cardiac physical exam. His teaching sessions using the bedside exam and heart sounds simulator were among the most cherished experiences of many residents and fellows, and the basis upon which many trainees selected careers in pediatric cardiology. He was the recipient of several "best teacher" awards. Furthermore, he was responsible for re-establishment of the California Society of Pediatric Cardiology, which more recently became the Western Society of Pediatric Cardiology, one of the largest and most prestigious pediatric sub-specialty meetings in the country.

Dr. Arno Hohn (left) with colleagues at CHLA.

Dr. Arno Hohn (center) with graduating fellows, Drs. Anjan Batra (left) and William Castillo (right).

Left-to-right: Drs. Robert Stanton, Anjan Batra and Arno Hohn.

Amongst all who had the pleasure of his acquaintance, Arno was beloved as a true gentleman who treated everyone with fairness and dignity. He was always considerate and kind, generous with his time, ever humble, and a man of his word.

Aside from his professional career, Arno enjoyed a very active social life with his wife Marie, three sons and many good friends. Arno loved the outdoors, and was an avid hiker and skier. He enjoyed second homes in Mammoth, California and later, in Vancouver, British Columbia, which provided him many years to continue his outdoor adventures.

In closing, he will be missed by his many colleagues and friends both within and outside Children's Hospital Los Angeles.

CCT

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Highlights of the 2014 Specialty Review in Pediatric Cardiology Course

By Maria Serratto, MD

In the mid-seventies, as I started to prepare for the Pediatric Cardiology Board examination, I realized, to my astonishment, that there was no organized board review course to help candidates in this task. Thus, candidates had to rely on their own efforts to review the subject matter in preparation for the board.

After successfully passing the examination, I decided to organize a preparatory course covering all aspects of the specialty, taught by a distinguished faculty. Thus, the *Chicago Specialty Review in Pediatric Cardiology Board Review - CME Course* was born. The first course was offered in 1976 under the auspices of the Cook County Graduate School of Medicine of Chicago. That year, and for several courses to follow, the program was 2 days in length, attended by about 30 registrants from all parts of the United States. Over the years the course expanded to the present duration of 5 days, and since 2010, the course has been sponsored by the Section on Cardiology and Cardiac Surgery of the American Academy of Pediatrics (www.aap.org) in collaboration with the Society of Pediatric Cardiology Training Programs Directors. Our course audience has expanded over the years as well, with approximately 150 attendees from the US and abroad participating in the last offering.

Continuing advances in our specialty have no doubt contributed to this growth, particularly in recent years as practicing specialists already board-certified, prepare to meet recertification requirements and practitioners in general strive to remain current in our ever-expanding field.

The past 38 years have been an exciting journey both for me as the founding director, and for our dedicated faculties as we had the opportunity to come to know hundreds of fine young future specialists, and watch them advance in their careers.

We are especially proud to count a number of current pediatric cardiology department heads among our alumni.

This year, the 19th edition of the course, will be held in Chicago September 15th-19th, 2014 at the Holiday Inn Mart Plaza Hotel, located in the heart of downtown Chicago overlooking the river. It promises to be another great educational experience with 20 faculty members joining me to provide a content-filled 5 days. Whether you are participating for the first time, or you are a past course participant, we would be honored to have you with us in Chicago this September, an ideal month to enjoy the many amenities Chicago offers with its cultural, culinary and architectural treasures.

"Over the years the course expanded to the present duration of 5 days...."

Details about the 2014 course are available at: www.aap.org/Pediatric-Cardiology-2014.

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Image of the Month #10: June, 2014 - The Archiving Working Group

Contributors: Jorge M. Giroud, MD; Robert Anderson, MD; Vera D. Aiello, MD; Diane E. Spicer, BS; Charles W. Shepard, MD; Jeffrey P. Jacobs, MD

AWG Web Portal Link for this Series of Images

http://www.accd-awg.umn.edu/Coronary_Disease/ALCAPA_09_41_03/ALCAPA_09_41_03_RPA.html

http://www.accd-awg.umn.edu/Coronary_Disease/ALCAPA_09_41_03/ALCAPA_09_41_03_LPA.html

IPCCC:

1. Anomalous pulmonary origin of the left coronary artery from the right pulmonary artery: 09.41.03, Q1.45.84
2. Anomalous pulmonary origin of the left coronary artery from the left pulmonary artery: 09.41.03, Q1.45.85

AEPC Derived Term:

1. Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) - coronary arterial origin from right pulmonary artery (09.41.03, Q1.45.84) or Anomalous origin of coronary artery from right pulmonary artery (09.46.04)
2. Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) - coronary arterial origin from left pulmonary artery (09.41.03, Q1.45.85) or Anomalous origin of coronary artery from left pulmonary artery (09.46.05)

EACTS-STS Derived Term:

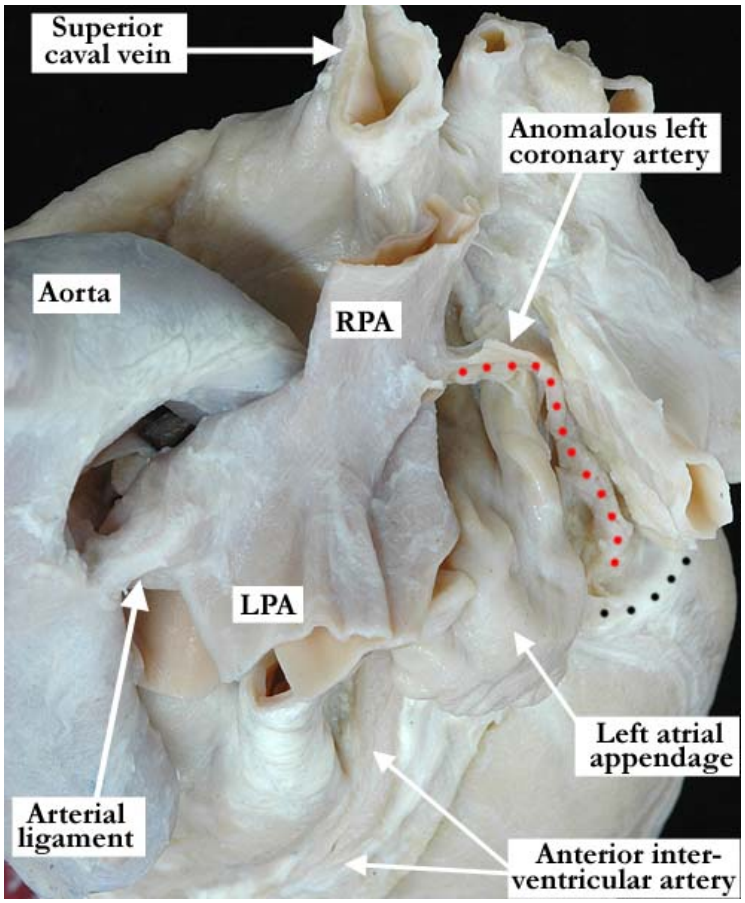
1. Coronary anomaly, APOC (Anomalous pulmonary origin of coronary), ALCAPA (Anomalous left main coronary artery from PA), Origin from right pulmonary artery (09.41.03, Q1.45.84)
2. Coronary anomaly, APOC (Anomalous pulmonary origin of coronary), ALCAPA (Anomalous left main coronary artery from PA), Origin from left pulmonary artery (09.41.03, Q1.45.85)

ICD10 Derived Term:

Malformation of coronary vessels (Q24.5)

Please visit us at the AWG Web Portal at http://ipccc-awg.net/ and help in the efforts of the Archiving Working Group and the International Society for Nomenclature of Paediatric and Congenital Heart Disease.

The authors would like to acknowledge the Children's Heart Foundation (http://www.childrensheartfoundation.org/) for financial support of the AWG Web Portal.

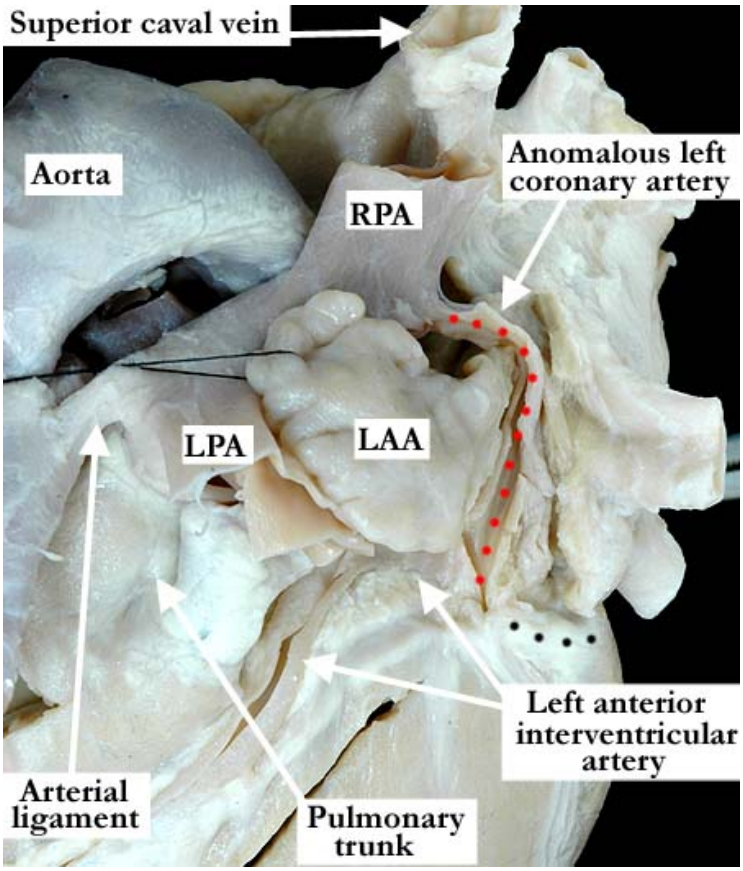


Left coronary artery from the right pulmonary artery:

Orientation: Posterior lateral view
Description: In this posterior lateral view the aorta has been retracted anteriorly to show the bifurcation of the right (RPA) and left (LPA) pulmonary arteries. There is an anomalous origin of the left coronary artery (red dots) from the right pulmonary artery. It extends over the left atrial appendage and bifurcates into the left anterior interventricular and circumflex (black dots) branches once it reaches the epicardial surface. Although not shown, the right coronary artery originates from the aorta.

Commentary

As discussed in the 9th Image of the Month Column published in the April 2014 issue of *Congenital Cardiology Today,* anomalous pulmonary origin of the left coronary artery is a rare congenital defect. The most common origin of the anomalous left coronary artery is from one of the sinuses of the pulmonary trunk, typically the left-handed sinus. Much less common is origin of the anomalous coronary artery from either branch of the pulmonary

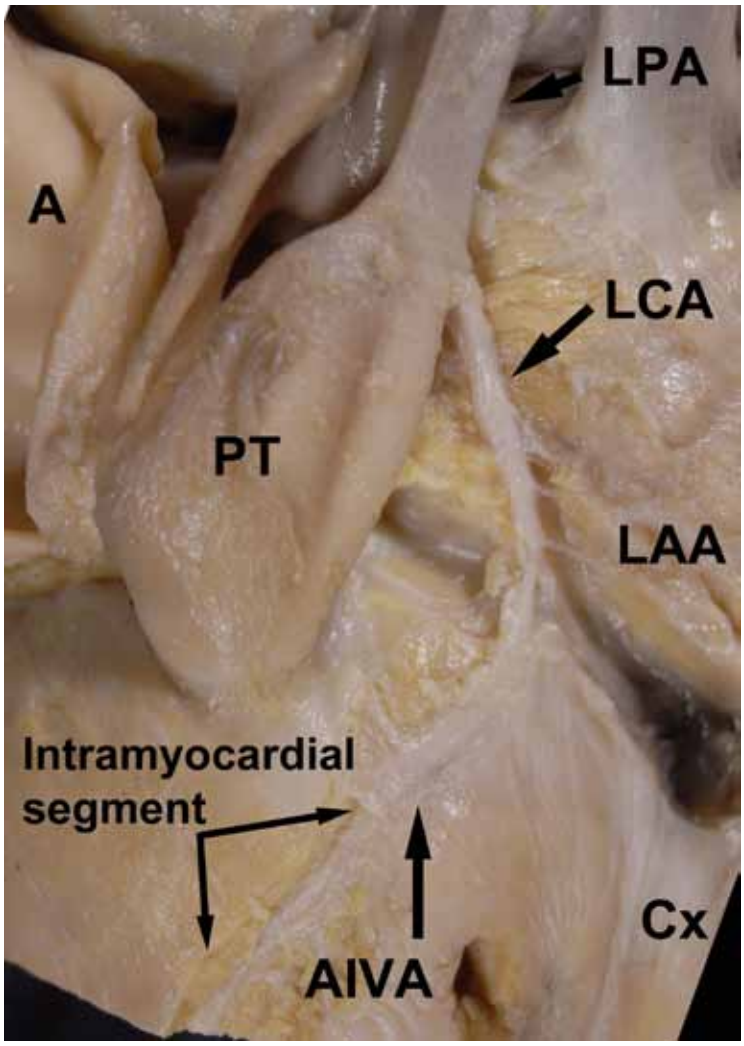


Left coronary artery from the right pulmonary artery:

Orientation: Posterior lateral view
Description: In this posterior lateral view the left atrial appendage (LAA) has been retracted better to show the bifurcation of the right (RPA) and left (LPA) pulmonary arteries. There is an anomalous origin of the left coronary artery (red dots) from the right pulmonary artery. It extends over the left atrial appendage and bifurcates into the left anterior interventricular and circumflex (black dots) branches once it reaches the epicardial surface.

trunk. In this column, we present the findings from two separate specimens showing the unusual anomalous origin of the left coronary artery from the right and left pulmonary arterial branches.

"The most common origin of the anomalous left coronary artery is from one of the sinuses of the pulmonary trunk, typically the left-handed sinus."



Left coronary artery from the left pulmonary artery:

Orientation: Left lateral superior view
Description: This image illustrates the anomalous pulmonary origin of the left coronary artery (LCA) from the proximal segment of the left pulmonary artery (LPA) as it leaves the pulmonary trunk (PT). The long 'trunk' of the left coronary artery gives rise to the circumflex (Cx) and anterior interventricular (AIVA = LAD) arteries. The left coronary artery has a short intramyocardial course in the atrial myocardium close to the origin of the left atrial appendage (LAA). The anterior interventricular artery also has a long and deep myocardial bridge. Although not illustrated, this patient also had a right-sided aortic arch (A) with mirror-image origin of the arch arteries and also a lung sequestration (right lower lobe) with separate arterial supply and venous drainage.

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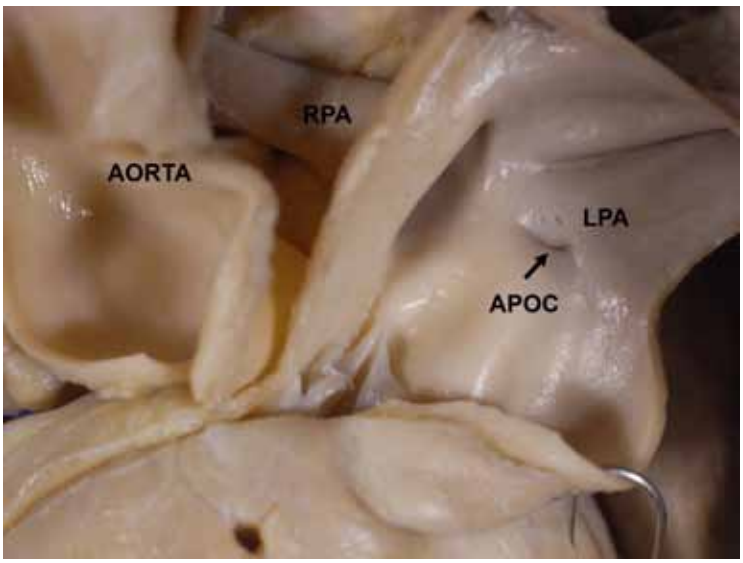
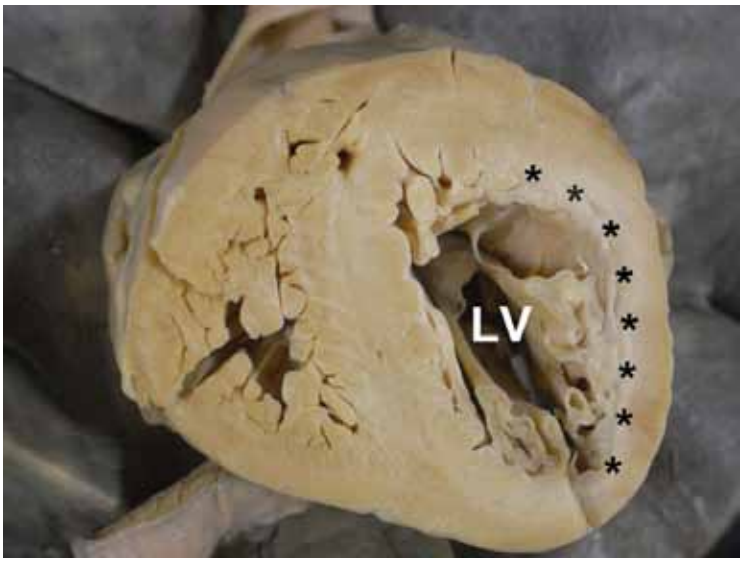


Left coronary artery from the left pulmonary artery:

Orientation: View of the opened great arteries
Description: This image illustrates the anomalous pulmonary origin of the left coronary artery from the proximal segment of the left pulmonary artery (LPA) as viewed from inside the left pulmonary artery. The left coronary orifice is labeled APOC, standing for anomalous pulmonary origin of the coronary artery (RPA - right pulmonary artery).



Left coronary artery from the left pulmonary artery:

Orientation: Cross sectional view of the ventricles from the apex
Description: This image of the ventricles viewed in cross section from the apex, shows the ischemic myocardium (marked by the asterisks) in a patient with anomalous origin of the left coronary artery from the pulmonary trunk. It involves mainly the supero-posterior free wall of the left ventricle (LV - left ventricle).

The authors would like to acknowledge the Children's Heart Foundation (www.childrensheartfoundation.org/) for financial support of the AWG Web Portal.

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Digisonics Introduces SMART for Streamlined Pediatric Reporting Workflow

Digisonics has introduced a new feature for its Cardiovascular Information System (CVIS) Solutions called SMART, Structured Macros and Report Templates. Developed specifically to cater to the pediatric reporting workflow, SMART provides users with the convenience of quickly creating a report template through the use of comments.

Going through the list of comments, users quickly build their report templates with just a few clicks instead of working through multiple forms. In addition, databased fields (such as study measurements) are autopopulated directly into the comments and templates providing enhanced workflow efficiencies. The SMART feature comes loaded with a comprehensive list of highly configurable congenital macros/templates. SMART provides a high level of user configurability while at the same time, not compromising the benefits of a structured report.

Digisonics provides top-rated clinical image management and structured reporting systems for cardiovascular (CVIS), radiology, and obstetrics & gynecology. Digisonics' structured reporting solutions combine high performance image review workstations, a powerful PACS image archive, an integrated clinical database, comprehensive analysis capabilities and highly configurable reporting for multiple modalities. Key applications are complemented with interfaces to information systems and 3rd party vendors, providing facilities with a seamless, efficient clinical workflow. To view a sample pediatric cardiography report go to: www.congenitalcardiology.com/SamplePediatricEchoReport.pdf.

For further information: www.digisonics.com.

Study Evaluates Role of Infliximab in Treating Kawasaki Disease: Antibody Treatment Helps Children with Dangerous Heart Disorder

Newswise — Kawasaki Disease (KD) is a severe childhood disease that many parents, even some doctors, mistake for an inconsequential viral infection. If not diagnosed or treated in time, it can lead to irreversible heart damage.

Signs of KD include prolonged fever associated with rash, red eyes, mouth, lips and tongue, and swollen hands and feet with peeling skin. The disease causes damage to the coronary arteries in a quarter of untreated children and may lead to serious heart problems in early adulthood. There is no diagnostic test for Kawasaki Disease, and current treatment fails to prevent coronary artery damage in at least one in 10 to 20 children and death in one in 1,000 children.

Between 10% - 20% of patients with KD experience fever relapse following the standard therapy with a single infusion of intravenous immunoglobulin (IVIG) and aspirin. It is known that IVIG resistance increases the risk of heart damage, most commonly a ballooning of the coronary arteries called aneurysms. These children require additional therapy to interrupt the inflammatory process that can lead to damage of the coronary arteries.

A study led by physicians at the University of California, San Diego School of Medicine and Rady Children's Hospital-San Diego looked at intensification of initial therapy for all children with KD in order to prevent IVIG-resistance and associated coronary artery abnormalities by assessing the addition of the medication infliximab to current standard therapy. The results of their study were published in the February 24, 2014 online issue of the medical journal *Lancet*.

Tumor necrosis factor &alpha (TNF&alpha) is a molecule made by the body that plays a role in the development of inflammation in KD; therefore, treatment with a TNFa antagonist is a logical therapeutic intervention, according to the researchers. Early experience with infliximab – a monoclonal antibody that binds TNFa – showed promising results. A Phase 1 trial in children with KD and persistent fever following standard therapy found no infusion reactions or serious adverse events, and subsequent studies suggested that infliximab led to faster resolution of fever and fewer days of hospitalization than a second IVIG infusion.

The UC San Diego researchers conducted a trial of 196 subjects at two centers – Rady Children's Hospital-San Diego, a research affiliate of UC San Diego School of Medicine, and Nationwide Children's Hospital in Columbus, Ohio – to assess whether infliximab could reduce IVIG treatment resistance.

"While the addition of infliximab to primary treatment in acute KD did not reduce treatment resistance, it was safe and well-tolerated, achieved a greater reduction in the size of the left coronary artery, and reduced the number of days of fever and laboratory markers of inflammation," said the study's first author, Adriana H. Tremoulet, MD of the UC San Diego Department of Pediatrics and the UC San Diego/Rady Children's Hospital-San Diego Kawasaki Disease Research Center. "We conclude that use of infliximab is safe in infants and children and that early treatment could help children with Kawasaki Disease with high levels of inflammation or early signs of coronary artery damage."

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