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Transcatheter Perforation of Atretic Pulmonary Valve in Pulmonary Atresia with Ventricular Septal Defect

By P. Syamasundar Rao, MD

Introduction

Transcatheter perforation of atretic pulmonary valve membrane by blunt end of regular[1-7] or coronary[8-10] guide wires, laser wires,[11-13] or radiofrequency catheters/ wires[3,4,13-22] followed by balloon dilatation has been used as an alternative to surgery in the management of pulmonary atresia with intact ventricular septum, with reasonably good results. However, such an approach to treat pulmonary atresia with Ventricular Septal Defect (VSD) has rarely been used.[12,23-25] This difference may in part be related to anatomic differences between the two types of pulmonary atresia. The purpose of this communication is to present the details of retrograde perforation of atretic pulmonary valve membrane along with anterograde balloon pulmonary valvuloplasty in a patient with pulmonary atresia with VSD (severe Tetralogy of Fallot) and to discuss the role of these procedures in the management of such cases.

"Transcatheter perforation of atretic pulmonary valve membrane by blunt end of regular[1-7] or coronary[8-10] guide-wires, laser wires,[11-13] or radiofrequency catheters/ wires[3,4,13-22] followed by balloon dilatation has been used as an alternative to surgery in the management of pulmonary atresia with intact ventricular septum, with reasonably good results. However, such an approach to treat pulmonary atresia with Ventricular Septal Defect (VSD) has rarely been used.[12,23-25]"

Case Report

A three-week-old female infant presented with severe cyanosis (in 1995) and, with cardiac catheterization and selective cine-angiography, was found to have a large VSD and pulmonary valve atresia and underwent urgent central aortopulmonary shunt. The infant improved clinically, and was discharged home shortly after surgery, and followed periodically in the outpatient clinic. Because of increasing cyanosis and polycythemia, she was referred back to the author for catheter evaluation at the age of

Table 1. Cardiac Catheterization Data Prior to and Immediately after Pulmonary Valve Perforation and at One-Year Follow-up						
	Cardiac Cath - December 1997				Cardiac Cath - December 1998	
	Prior to PV Perforation		After PV Perforation			
Site	O_2 Sat, %	Pressure, mmHg	O_2 Sat, %	Pressure, mmHg	O_2 Sat, %	Pressure, mmHg
SVC	60		60		64	
IVC	62				70	
RA	60	m-4.5	60	m-5	64	m-7
RV	68	85/8		102/8	67	98/12
MPA	77	25/12; m-18	83	35/15; m-20	88	40/12; m-19 LPA-26/16;m-18
LA	96	m-8			97	m-13
LV	96	85/8		102/8	97	98/12
Ao	77	85/40; m-62	87	102/60; m-80	90	98/65;m-80
Qp:Qs	1.1:1.0		1.3:1.0		2.3	
PVR	1.6		2.0		0.5	

Ao, aorta; Cath, catheterization; IVC, inferior vena cava; LA, left atrium; LV, left ventricle; LPA, left pulmonary artery; m, mean; MPA, main pulmonary artery; PV, pulmonary valve; PVR, pulmonary vascular resistance; Qp:Qs, pulmonary to systemic flow ratio; RV, right ventricle; SVC, superior vena cava.

22 months. Right and left heart catheterization (Table 1) with selective cine-angiography confirmed the diagnosis of a large malaligned VSD with bidirectional shunt, pulmonary valve atresia and patent aortopulmonary shunt. There was moderate arterial desaturation (77%) with a pulmonary to systemic flow ratio (Qp:Qs) of 1.1 and normal pulmonary vascular resistance (Table 1).

The shunt was cannulated from the aorta with #4-F Glidecath (Meditech, Inc Watertown, MA) with the help of Benston guide-wire (Cook, Bloomington, IN) and a pulmonary artery (PA) cineangiogram was performed to delineate the PA anatomy (Figure 1a). Simultaneous injection of the contrast material into the main PA and right ventricular outflow tract was also performed to visualize the atretic membranous pulmonary valve (Figure 2). Brief attempts to position a catheter below the pulmonary valve anterogradely and to perforate the valve were unsuccessful. Therefore, a retrograde approach was entertained. The #4-F Glidecath catheter that was positioned in the PA via the central shunt was connected to a Toughy-adapter to facilitate injection of the contrast material over the wire. A 0.021 inch straight guide wire was positioned at the tip of the catheter and after assuring that the catheter was in the middle of the pulmonary valve by a test injection through the Toughy-adapter, firm, but gentle pressure was applied, thus allowing the guide wire to traverse through the atretic pulmonary valve. Over this guide wire, the catheter was advanced into the right ventricle and then the guide wire and the catheter together were advanced into the right atrium. At this juncture the guide wire was exchanged with an exchange length 0.025 inch "J" tipped extra stiff Amplatz guide wire (Cordis Corporation, Miami, FL). The "J" component of the guide wire in the right atrium was snared with a 15mm "gooseneck" snare (Microvena, Vadnais Heights, MN) that was introduced through the femoral vein. The tip of the wire was slowly drawn out through the femoral vein while slowly advancing it from the femoral artery entry. Thus, a loop was formed from the femoral artery through descending and ascending aorta, aortopulmonary shunt, main pulmonary artery, perforated pulmonary valve, right ventricle, right atrium, inferior vena cava, and femoral vein. The size of the pulmonary valve annulus measured 6 to 7 mm.

A 6mm diameter, 2cm long Ultrathin balloon angioplasty catheter (Meditech) was introduced over the femoral venous end of the guide wire and was advanced

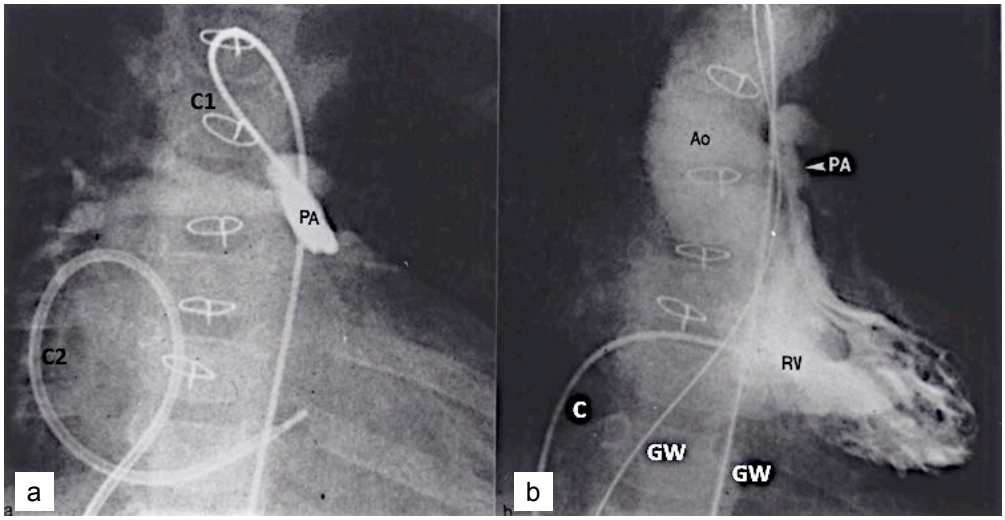
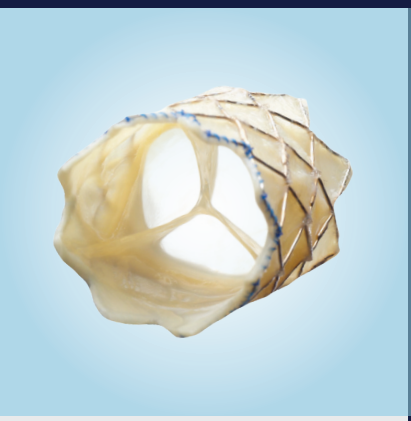


Figure 1a. Selected cine-angiographic frame of main pulmonary artery (PA) in posteroanterior view prior to pulmonary valve perforation demonstrating blindly ending PA with opacification of the branch PAs (not labeled). The catheter (C1) was introduced into the PA from the aorta via the central aortopulmonary shunt. C2, catheter in the right atrium. 1b. Selected cine-angiographic frame of right ventricle (RV) in postero-anterior view following pulmonary valve perforation demonstrating forward flow from the RV into the PA. Components of the guide wire (GW) rail are marked. Ao, aorta; C, catheter in the RV.

RIGHT DATA.



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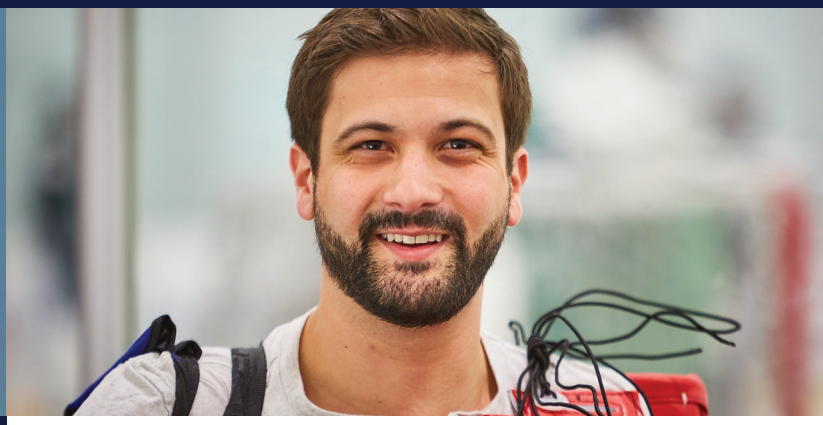

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*The term "stent fracture" refers to the fracturing of the Melody TPV. However, in subjects with multiple stents in the RVOT it is difficult to definitively attribute stent fractures to the Melody frame versus another stent.

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- Implantation of the TPV in the left heart
- RVOT unfavorable for good stent anchorage
- Severe RVOT obstruction, which cannot be dilated by balloon
- Obstruction of the central veins
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- Active endocarditis
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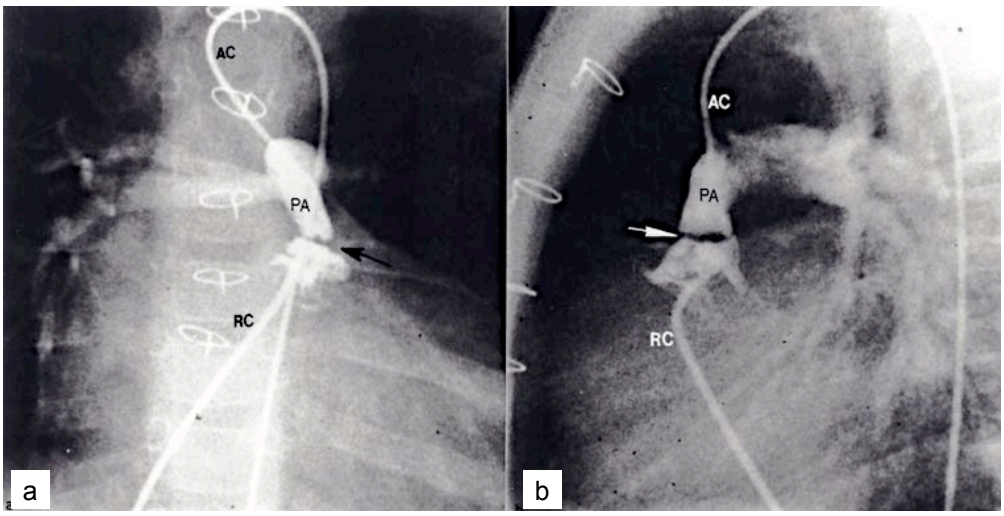

Figure 2. Selected cineangiographic frames in posteroanterior (a) and lateral (b) projections of simultaneous injection of the contrast material into the main pulmonary artery (PA) via a catheter introduced through the central aortopulmonary shunt (AC) and right ventricular outflow tract via a catheter (RC) introduced from the right atrium demonstrating atretic membranous pulmonary valve (black arrowhead in a and white arrowhead in b).

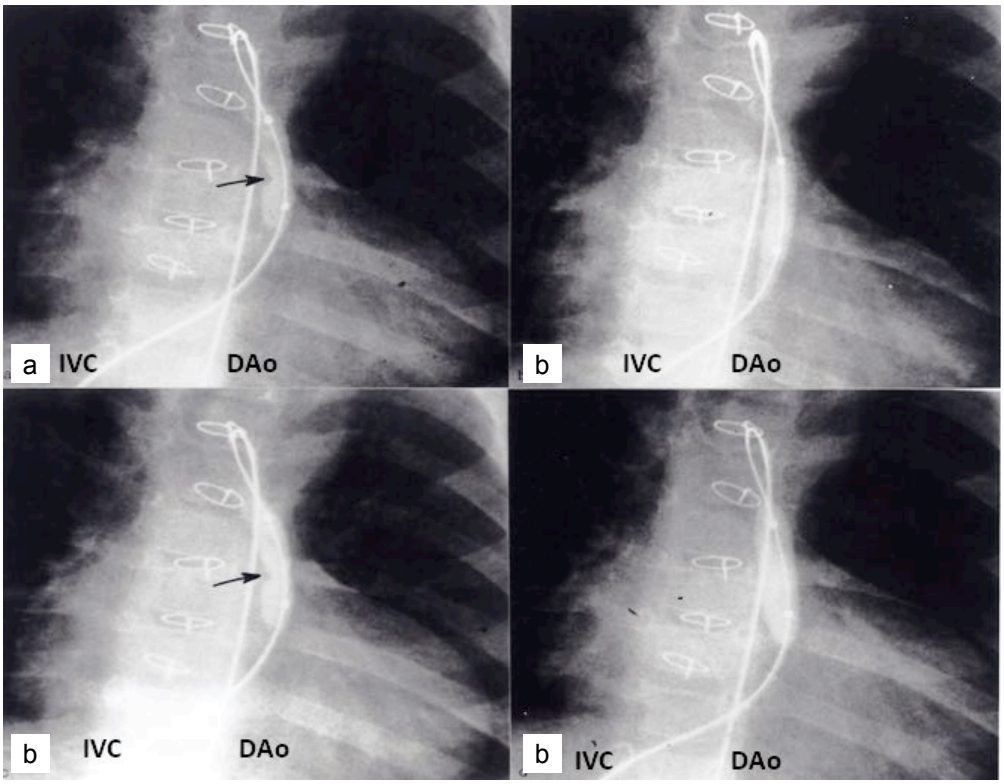

Figure 3. Selected cine-radiographic frames in posteroanterior view showing balloon angioplasty catheters introduced over the femoral venous end of the guide wire and positioned across the atretic but perforated pulmonary valve. Waisting (arrow in a) of the 6 mm balloon is shown which disappeared on further balloon inflation (b). Similar waisting (arrow in c) of the 8 mm balloon is shown which also disappeared on further balloon inflation (d). DAo, descending aorta; IVC, inferior vena cava.

and positioned across the atretic, but perforated, pulmonary valve. Two balloon inflations were performed with resultant ablation of the waist of the balloon (Figures 3a and 3b). The balloon was deflated and exchanged with an 8mm, 2cm long Ultrathin Meditech balloon catheter and three balloon inflations were performed (Figures 3c and 3d). The balloon angioplasty catheter was removed, while leaving the guide wire still in place. A right ventricular cine-angiogram was performed (Figure 1b) demonstrating flow across the opened pulmonary valve. The Glidecath catheter from the aorta was advanced over the guide-wire loop into the right atrium and the Amplatz guide was removed from the arterial site. This was performed so that the withdrawal of the guide wire does not injure intra-cardiac structures. Subsequent to the removal of the guide wire, careful pressure pullback tracings from the right atrium, right ventricle, pulmonary artery, aortopulmonary shunt and aorta were performed, and aortic saturation, as well as superior vena caval saturation and pulmonary arterial saturation were repeated as were the pressures (Table 1). No complications were encountered during or after the procedure. Overnight observation using pulse oxymetry revealed O_2 saturations in mid to high 80's. Echo-Doppler study on the morning following the procedure demonstrated forward flow across the right ventricular outflow tract and pulmonary valve, similar to angiography as shown in Figure 1b.

In summary, the procedure of guide-wire perforation followed by balloon pulmonary valvuloplasty produced an increase in arterial oxygen saturation (77% vs. 87%) without increasing pulmonary vascular resistance (Table 1). It was envisioned that the extra flow produced by opening the pulmonary valve would result in growth of the pulmonary arteries, making the patient a lower-risk candidate for total surgical correction in the future.

One year following the procedure, a repeat cardiac catheterization was performed preparatory to total surgical correction. Catheterization data (Table 1) revealed good arterial O_2 saturation (90%) and normal pulmonary vascular resistance. Angiographic anatomy is similar to the prior study, with slight improvement in the size of the PA, but with persistence of the branch PA stenosis at their origin. The distal pulmonary arteries appeared normal. Based on these data, surgical correction with closure of the VSD, relief of right ventricular infundibular obstruction and enlargement of the main and origins of the branch pulmonary arteries was recommended and was successfully undertaken a few weeks after the cardiac catheterization.

Discussion

Opening of the pulmonary valve either by surgical[26,27] or transcatheter[1-22,28-31] approaches is the mainstay in the initial management of patients with pulmonary atresia with intact ventricular septum. We have used both the blunt end of guide wires (Figure 4) and radio frequency wires (Figures 5, 6 and 7) to accomplish this procedure. In contradistinction; aorto-pulmonary shunts are usually performed for patients with pulmonary atresia with VSD.[32-34] Subsequently, total correction with VSD closure and reconstruction of the right ventricular outflow tract is undertaken.[32-34] In the case presented, we took an unusual step to transcatheter open the pulmonary valve with the intent to encourage

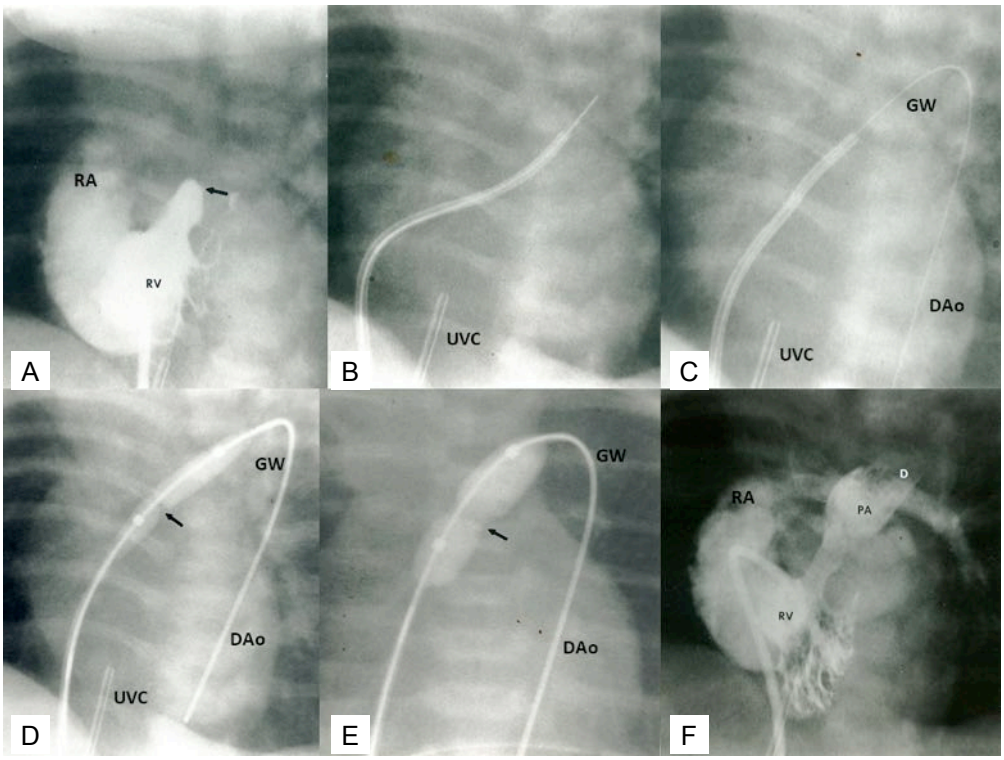

Figure 4. A. Selected cine frame from right ventricular (RV) angiogram in a sitting-up (150 LAO and 350 cranial) view demonstrating an atretic pulmonary valve (arrow). The right atrium (RA) is opacified due to tricuspid insufficiency. B & C. After perforation of the atretic pulmonary valve with the blunt end of a guide wire (GW), it was advanced into the pulmonary artery, ductus (not labeled) and then into the descending aorta (DAo). D & E. A five-mm diameter and then, an 8-mm diameter balloon dilatation catheters were positioned across the perforated pulmonary valve membrane, showing "waisting" of the balloon (arrows) during the initial phases of balloon inflation which has disappeared after complete inflation of the respective balloons (Not shown). F. Selected cine frame from RV angiogram at the conclusion of the procedure demonstrating opacification of the pulmonary artery (PA) and its branches. Also note opacification of pulmonary end of patent ductus arteriosus (D). There was significant tricuspid insufficiency both before and after the procedure. UVC, umbilical venous catheter.

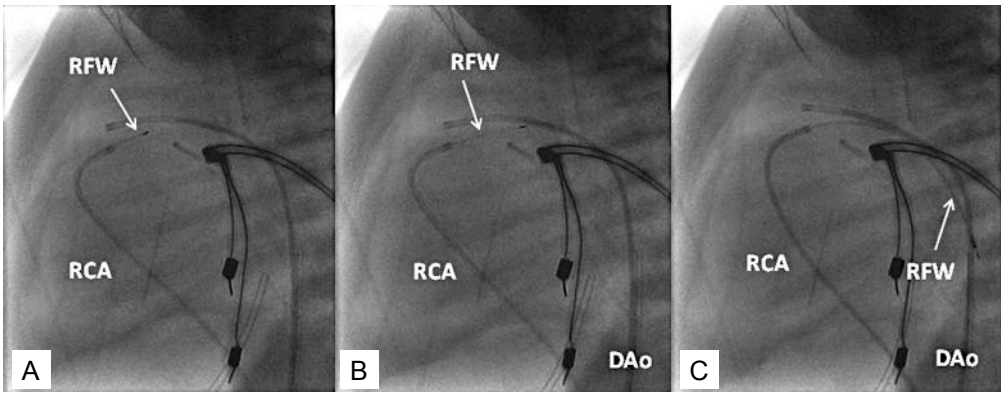

Figure 5. Selected cine-radiographic frames in straight lateral projections showing progressive advancement of the radiofrequency wire (RFW) (thin arrows) via a right coronary artery (RCA) catheter into the main pulmonary artery (A & B) and from there into the descending aorta (DAo) via the ductus arteriosus (not marked) (C).

growth of the PAs and reduce the risk of total surgical correction, given the state of surgical methods/results in late 1990s.

The differences in approach may in part be related to anatomic differences between the two types of pulmonary atresia. In pulmonary atresia with intact ventricular septum, the main and branch pulmonary arteries are usually well formed and near normal in caliber whereas in pulmonary atresia with VSD, the pulmonary arteries are small and hypoplastic in most patients making it difficult to adopt transcatheter approaches in their management. Presumably, for the reason, there are only a limited number of reports describing transcatheter perforation of the atretic pulmonary valve in patients with pulmonary atresia and VSD; extensive literature search (PubMed) revealed only a few such reports.[12,23-25] Qureshi and associates[12] percutaneously perforated atretic pulmonary valve in two children aged 2 years and 2 weeks, respectively, who had prior modified Blalock-Taussig shunts. Perforation of the pulmonary valve was undertaken anterogradely in one patient and retrogradely via the shunt in the other patient. Successful laser-wire perforation and balloon dilatation was performed in both patients, and right ventricle (RV) to PA continuity was established. During follow-up for two to six months, the O_2 saturations remain improved and both patients were thought to have become suitable candidates for total surgical correction without additional palliative surgery. Kuhn et al[23] performed perforation of atretic pulmonary valve with the stiff end of a 0.014" guide wire, introduced anterogradely in a 5-day-old infant with pulmonary atresia with VSD followed by balloon dilatation of the pulmonary valve. They were able to establish RV-to-PA continuity. At the age of four-months, they demonstrated growth of the pulmonary arteries and complete correction was planned in three to six months from that time. The authors recommended this procedure to encourage growth of the PAs as an alternative to surgery, but failed to acknowledge prior report[12] of percutaneous perforation of pulmonary valve in patients with pulmonary atresia with VSD. Walsh and colleagues[24] employed radiofrequency perforation of the pulmonary valve membrane as a palliative strategy for pulmonary atresia with VSD; they were successful in performing the procedure in six of the eight children, establishing RV-to-PA continuity. Three of these children had complete correction within one year of the procedure without requiring additional palliative procedures and others await further surgery. Khan and associates[25] perforated the pulmonary valve with a radiofrequency wire in an adult patient with Tetralogy of Fallot and pulmonary atresia who had prior palliative surgical procedures in infancy and childhood. They used a Nykanen radiofrequency perforation catheter (Baylis Medical Company Inc, Montreal, Canada), followed by dilatation of the pulmonary valve with an 8 mm balloon dilatation catheter. The O_2 saturations improved to 85% and RV-to-PA continuity was established. Follow-up at three months revealed continued clinical improvement. They planned to re-evaluate the patient in 6 to 12 months time to consider a more definite repair. Some interventionalists have used a hybrid perventricular approach to perforate pulmonary valve along with RV outflow tract stent placement to palliate low-weight infants with pulmonary atresia with VSD.[35,36]

Conclusion

Though there is limited experience, percutaneous perforation of the pulmonary

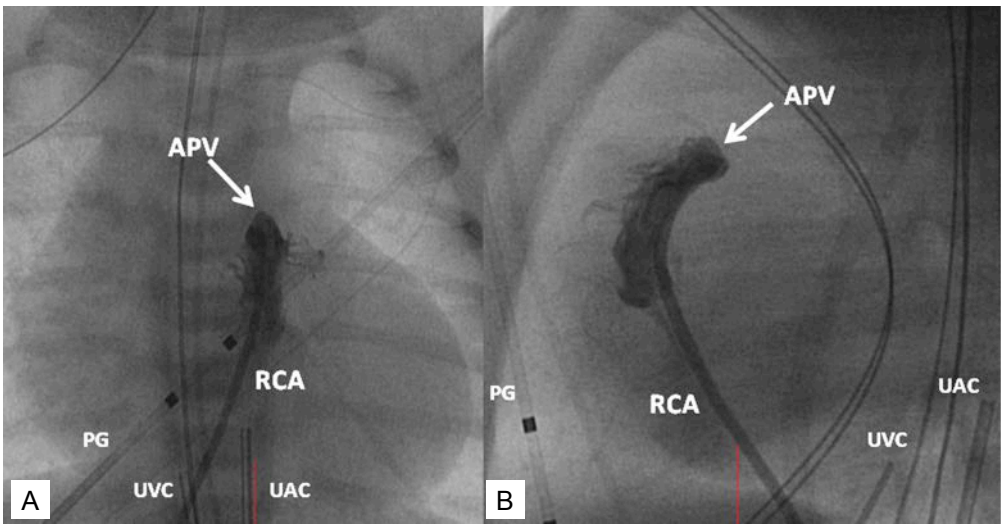

Figure 6. Selected cineangiographic frames from right ventricular outflow tract cine in postero-anterior (A) and lateral (B) views in a neonate with pulmonary atresia with intact ventricular septum showing the position of the tip of the right coronary artery (RCA) catheter in close proximity to the atretic pulmonary valve (APV) (arrows in A and B). PG, pigtail catheter placed on the baby's body for calibration purposes; UAC, umbilical artery catheter; UVC, umbilical venous catheter.

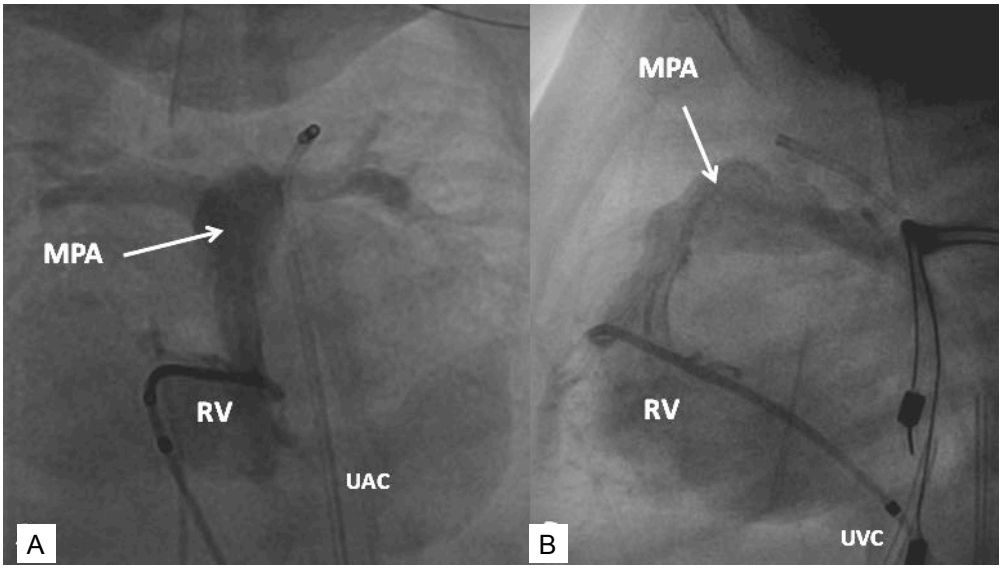

Figure 7. Selected cine-angiographic frame from right ventricular (RV) cine in a sitting up (150 LAO and 350 cranial) (A) and straight lateral (B) views immediately after radiofrequency perforation and balloon pulmonary valvuloplasty demonstrating prompt opacification of the main pulmonary artery (MPA). This is the same infant shown in Figure 6. UAC, umbilical artery catheter; UVC, umbilical venous catheter.

valve (followed by balloon dilatation) as an initial palliative procedure to augment pulmonary blood flow seems to be an effective alternative to palliate surgery in patients with pulmonary atresia with VSD, and such establishment of RV-to-PA continuity may result in better surgical outcomes after eventual corrective surgery.

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Mobil Cath Lab for the Treatment of Congenital Heart Defects - Is It a Feasible Idea? My Own Experience and Example of Rwanda

By Jacek Bialkowski, MD

Our interest in international collaboration and development of Paediatric Cardiology, as well as interventional procedures in Congenital Heart Defects (CHD), has existed for many years. Our team was educated mostly in the Spanish Language Centers (such as Instituto Nacional de Cardiologia Ignacio Chavez in Mexico DF or Hospital Ramon y Cajal in Madrid, Spain). Later, we invited Drs. Ramon Bermudez Canete, Luigi Ballerini, Chuck Mullins and others to our center in Poland to perform difficult procedures. In the years between 2004-2006, M. Szkutnik and I organized three workshops related to progress in non-surgical treatment of CHD in Hospital Obrero in La Paz (Bolivia). Those experiences we described in an issue of *Congenital Cardiology Toda*y in 2007.[1] We described that at high altitude (La Paz is situated about 3,800 meters above sea level) exist patients with Patent Ductus Arteriosus (PDA), mostly suitable for closure with an Amplatzer Duct Occluder Type I.[2] Moreover, some investigation was performed in a multicenter study, comparing PDAs in low lands (Poland, Spain) and high altitude (Mexico, Guatemala, Bolivia).[3] It was obvious that there are important differences in both populations: in the lowlands, PDAs had smaller diameter and pulmonary pressure was lower than in high altitude inhabitants. Since that time we started our educational program for young cardiologists in the transcatheter treatment of CHD. We obtained sponsors for fellowships from different institutions, like AGA Medical Corp. Occlutech Comp, Vaticane (The Pope John Paul II Grants) and Foundation for Children with Congenital Heart Defect in Zabrze, Poland. Finally, between 2007 and 2017, we trained twenty-two cardiologists (all below 40 years old) from different countries from Europe (Ukraine, Belarus, Bulgaria, Georgia, Russia, Italy), Latin America (Mexico, Bolivia, Guatemala, Argentina) and Asia (Uzbekistan, China) in our cath lab in Zabrze. The training period varied from one month to 12 months.

Since November 2011, when the Mother and Child Health Congress of WHO (World Health Organization) in Tashkent, Uzbekistan took place, we have been thinking about the feasibility of a mobile catheter laboratory mounted on a truck for Africa (Figures 1 and 2). While we have had support from key personnel at WHO, unfortunately, it did not progress further. Many other people and institutions were asked to help solve this problem. That too was unsuccessful. In 2014, during the *Pediatric Interventional Cardiac Symposium* in Chicago, Dr. Shak Qureshi (GB), past AEPC President, showed an interest in this idea. According to him, the Mobile Cath Lab might be defined and organized as follows:

What Is It: A mobile cath lab should consist of a specifically made cath lab mounted on a mobile transport truck. The catheter laboratory should consist of a single plane fluoroscopy machine with a floor-mounted table and a series of monitors for viewing the images.

The Catheter Laboratory Room: This will need to consist of a scrub area, an area where sterile gowns and gloves can be stored, and an area for two operators to scrub and prepare themselves. There would need to be a control room where the various personnel can aid in the procedures. Anaesthetic equipment would need to be located at the head end of the table, as well as a source for electricity and anaesthetic gases. There would also need to a safe and secure electrical and water supply.

The Truck: The truck would need to be of sufficient size to accommodate all of the above. In Africa, the roads may not always be tarmac when moving between cities and countries. The entire lab would need to be shock-proof to tolerate bumps on the roads, to prevent damage to the machines and the systems.

The Teams: For each mission, teams consisting of two interventional paediatric cardiologists, anaesthetists, anaesthetic assistants, a radiographer, a haemo-dynamic technician, two nurse assistants, three recovery nurses would be needed. Their living accommodations, food and other needs would need to be planned and understood well in advance.

Countries: The countries which have an interest in such a facility should be ones which have a significant need for the treatment of CHD, that are not being met locally by the institutions or by the governments or by other non-governmental organizations (NGOs). The governments and UNHCR (UN Refugee Agency) would need to give a commitment to support such a project along with the development of the infrastructure required.

Links with Local Hospitals: A local team, which needs training in the treatment of CHD, is essential for the longer term sustainability of a

Figure 1. Example of mobil cath lab on a truck trailer.

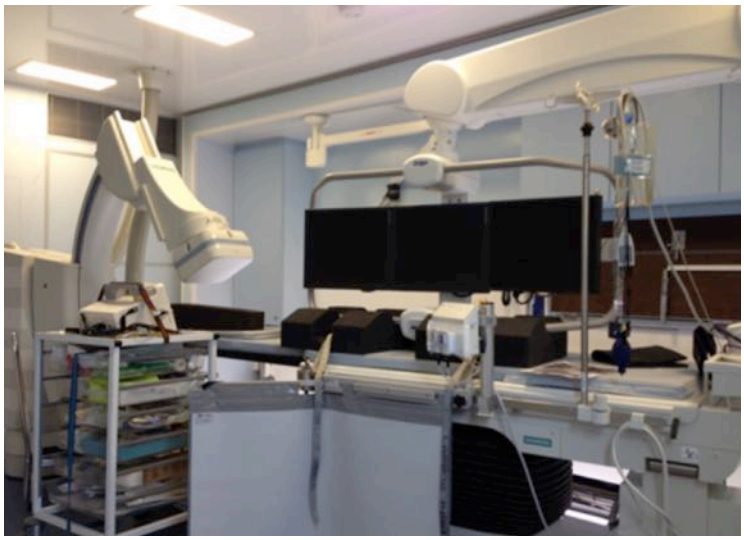

Figure 2. Mobil cath lab inside.

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Top: (L-R) Allen Tower Jr., President of NuMED with Dr. Ziyad Hijazi and Dr. Shakeel Qureshi.
Center: Dr. Ziyad Hijazi.
Bottom: (L-R) Dr. Michael Tynan, Allen Tower Sr., founder of NuMED and Dr. Shakeel Qureshi (PICS 2003).

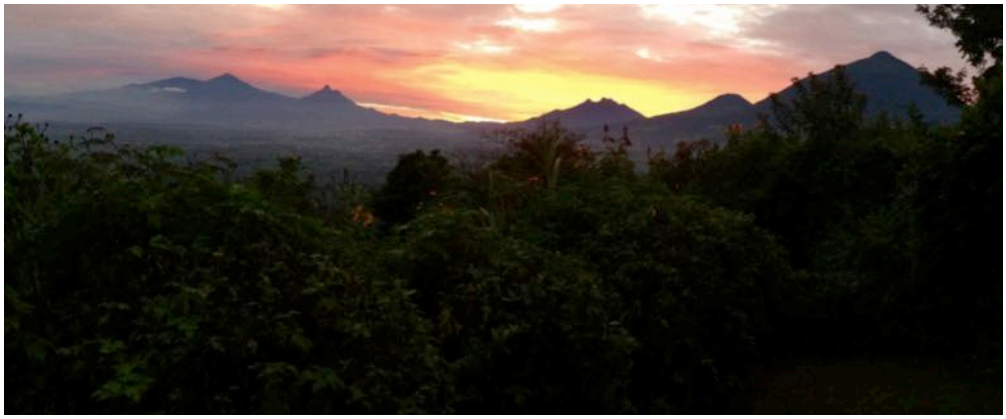

Figure 3. Rwanda, beautiful country of thousands. of hills.

Figure 4. Children awaiting construction of the new outpatient clinic.

Figure 5. Everything is possible in Rwanda.

Congenital Heart Disease interventional programme. The teams should consist of Paediatric Cardiologists, nurses and technicians, although technicians are not vital.

Links with Different Governments: In order to facilitate the work of the teams, and the movement of the mobile catheter laboratory between different cities, co-ordination will be needed between the different countries. As interventions are performed, the equipment that has been used will need to be replaced, and may have to be flown in. Each country will need to facilitate, not just the movement of the mobile catheter laboratory, but also the interventional equipment needed for the procedures. Visas and other types of political support will be needed for the teams from different countries, and would need to be coordinated so that one team works in one country (2-3 weeks), and another team flies in to carry out the treatments. Equipment will be needed for the interventional procedures, but many medical companies will provide donations of the equipment. Figures 1 and 2 are examples of existing mobil cath labs.

I was invited by Drs. S. Qureshi and H. Sivert to present this idea during the *CSI Africa Symposium* in Addis Abbeba, Ethiopia. During this meeting, African Pediatric Cardiologists expressed a moderate interest in this idea. The presentation was repeated during the main congress, *CSI Symposium* in June 2016 in Frankfurt with the same effect. I started to think that something was wrong, and that the idea might be too general, and I should change my strategy.

The next step was my "tourist" visit to Rwanda in January 2018. Rwanda is a small African, equatorial country. it has only 26,000 square km, with a population of 12.5 million inhabitants with a stormy contemporary history. But now, it is stable politically and economically. Rwanda is beautiful with a mostly mountain landscape, and is called the Thousands Hills Country (Figure 3). The population is predominantly children (Figures 4, 5). During my stay, I wanted to have meetings with local Pediatric Cardiology specialists; it was a difficult task. Only with help of local people and the invaluable aid of Priest Leszek Czelusniak, a Polish missionary, and his collaborating nurse from the U.S., Stacy Thomlison, was I able to find and contact Dr. Joseph Mucumbitsi. Dr. Mucumbitsi explained to me that in all of Rwanda, there are no regular cardiac catheterization labs. This is not an extraordinary situation in Africa. To my knowledge, there are no cath labs in Niger, Chad, Congo, Zambia, Guinea, Zimbabwe, Madagaskar or Burundi (See Figure 6). In Rwanda, there were only a few charity organizations from Europe (the Chain of Hope), a few missions and some simply percutaneous interventions in CHD that were performed with X-ray C arm.[4] So after my return, I informed Drs. Shak Qureshi and Horst Sivert about the situation in Rwanda. I explained that it might be possible to organize permanent cath labs through outside organizations. Through personal communication with Dr. M. Swierad, I had received confirmation that Knights of Malta, a Catholic charity, was interested in the project.

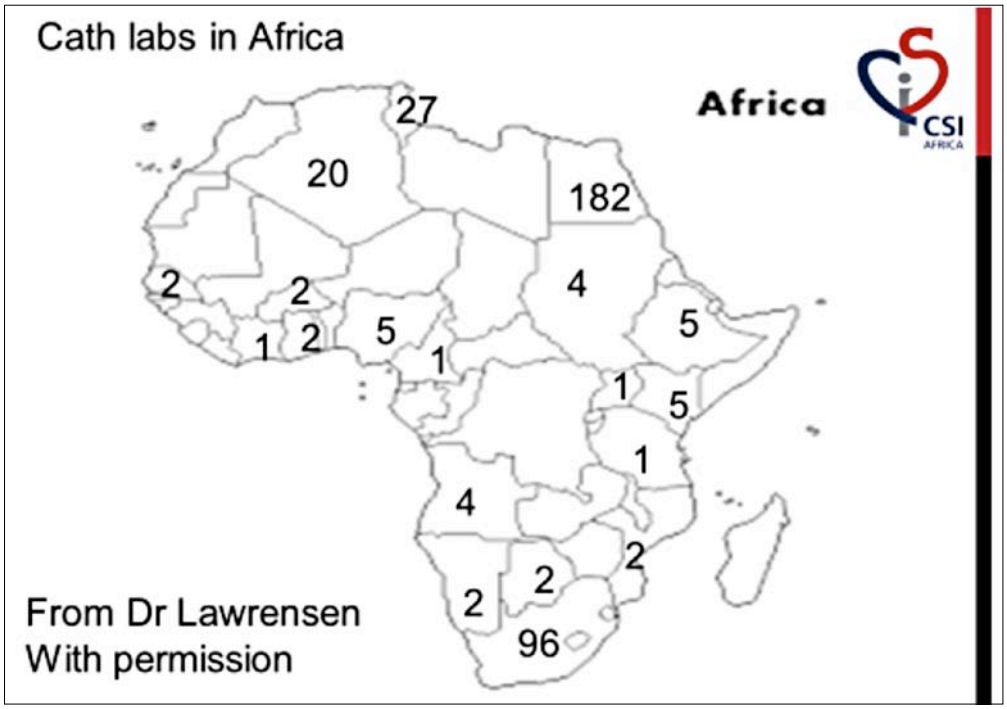

Figure 6. Locations of cath labs in Africa.

In the meantime, Rwanda was visited by Sir Magdi Yacoub from the Chain of Hope UK, and Prof. William Wijins from European Society of Cardiology. According to last mail I received from Dr. Mucumbitsi, Sir Yakoub wanted to built a center in Kigali, and they are working on a comprehensive five year plan.

So in conclusion, the idea of mobil cath labs for developing countries is not feasible. A more practical and useful plan is to organize fully-equipped, traditional cath labs, with financial, logistical and personnel support.

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Compiled and Reviewed by Kate Baldwin, Senior Editor Special Projects

Diagnostics of Genetic Cardiac Diseases using Stem Cell-Derived Cardiomyocytes

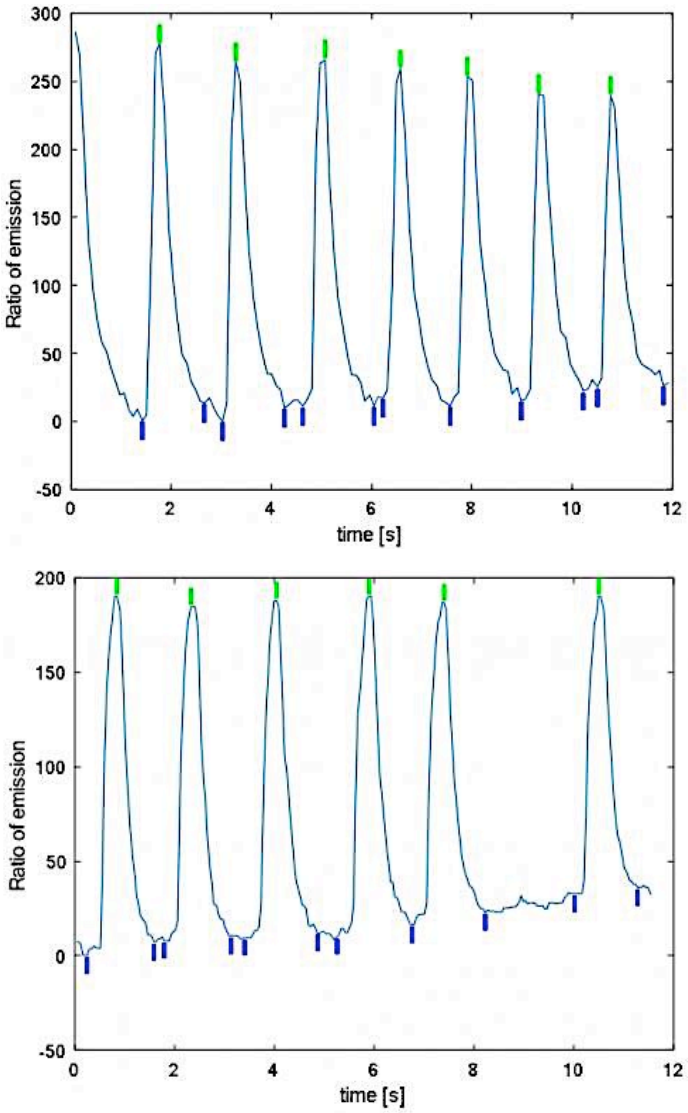

Figure 1(a) - Top. One signal of a stem cell originated from a healthy subject (b)- Bottom and the other from a CPVT patient.Credit: Authors.

A new study by Professors Martti Juhola and Katriina Aalto-Setälä of the University of Tampere in Finland demonstrates that with the use of artificial intelligence (AI) and machine learning, it is possible not only to accurately sort sick cardiac cell cultures from healthy ones, but also to differentiate between genetic cardiac diseases.

iPSC-derived cardiomyocytes can be derived from a blood sample or a skin biopsy. These cells are currently used to understand the pathophysiology of different diseases, and to identify new potential drugs for various diseases.

Machine learning and AI have greatly improved in recent years. Scientists at the University of Tampere have now combined stem cell technology and artificial intelligence to study beating cardiomyocytes in cell cultures. The beating behavior of the cells was analyzed using calcium signals. Calcium is essential for cardiomyocytes to beat, and the beating can be monitored by using fluorescent labels.

In the study, the cardiomyocytes were derived either from patients with a genetic arrhythmia (CPVT), Long QT Syndrome (LQTS), or hypertrophic cardiomyopathy (HCM), or from healthy individuals. The beatings of single cardiomyocytes were recorded, and the analysis software was taught what diseases they represented. The program then learned to separate the different groups, and to identify specific features in the beating behavior of each cell.

The software is now capable of identifying whether signals are from cells derived from an individual carrying a disease-causing mutation or from a healthy individual. This is very impressive, but the biggest surprise was that the program could also tell the difference between the diseases.

This important observation reveals that iPSC-derived cells and artificial intelligence have the potential to be used in diagnostics. Currently, genetic diseases are mainly diagnosed by DNA analysis, but in many cases the results do not reveal whether the DNA alteration is the true cause of the disease or whether it is just an innocent variation. This new finding demonstrates that uniting artificial intelligence and machine learning can help in such situations. The combination of technologies could also be used in cases of unspecific, but severe cardiac findings to identify the specific disease causing the symptoms.

See the article in *Scientific Reports:* www.nature.com/articles/s41598-018-27695-5

Poor Sleep Quality, Independent of Sleep Apnea, Is Established as an Important Risk Factor for Atrial Fibrillation

Poor sleep quality appears to be an important risk factor for Atrial Fibrillation (AF), report scientists in the first study of its kind to demonstrate a relationship between poor sleep quality independent of sleep apnea and a higher risk of Atrial Fibrillation. Their findings were published in *HeartRhythm.*

AF is an irregular, rapid heart rate that may cause symptoms, such as heart palpitations, fatigue, and shortness of breath. It

can substantially reduce quality of life, and is associated with heightened risks of stroke, dementia, heart attack, kidney disease, and death. Obstructive sleep apnea has been established as a risk factor for AF, but the mechanism is unclear. While episodes of abnormally slow or shallow breathing (hypopnea) and apnea may cause cardiopulmonary stress, induce inflammation, and contribute to cardiovascular disease, obstructive sleep apnea also results in poor sleep. Aspects of poor sleep, such as altered sleep duration, efficiency, and architecture have been linked to other cardiovascular diseases.

"While a relationship between sleep apnea and AF has previously been demonstrated, the effect of sleep itself on AF risk has remained unknown," explained lead investigator Gregory M. Marcus, MD, MAS, Electrophysiology Section, Division of Cardiology, Department of Medicine, University of California, San Francisco, CA, USA. "Strategies to enhance sleep quality are different from those that focus on relieving airway obstruction, so it is important to understand the relationship between sleep itself and AF."

Investigators drew on four different studies to determine whether poor sleep itself is a risk factor for AF. First, they used the global, internet-based Health eHeart Study, and determined that individuals with more frequent nighttime awakenings while trying to sleep more often, carried a diagnosis of AF. They then validated these findings by using the NIH-funded Cardiovascular Health Study, a prospective cohort study, in which they found that individuals who reported more frequent nighttime awakenings at baseline exhibited a higher risk of developing AF both before and after adjustment for potential confounders.

Within a subset of these individuals who had undergone formal sleep studies, they found that less REM sleep, in particular, predicted future AF. Finally, in order to see if these findings were readily translatable to patients already seen in healthcare settings and recognized by their providers as having sleeping difficulty, they drew on the California Healthcare Cost and Utilization Project (HCUP), a set of medical records databases of all California residents age 21 or older who received care in a California ambulatory surgery unit, emergency department, or inpatient hospital unit between January 2005 and December 2009. Among several million people, the HCUP data confirmed that a diagnosis of insomnia predicted a diagnosis of AF both before and after adjustment for potential confounding effects.

These results provide more evidence that sleep quality is important to cardiovascular health and specifically to AF. Investigators determined that there was no evidence that sleep duration per se was a risk factor for AF. Instead, they consistently found sleep disruption to be an important risk factor. While the underlying mechanisms are still unknown, these findings may motivate novel ways to think about, and hence future research into factors that influence AF risk.

This is the first study to demonstrate a relationship between worse sleep quality independent of sleep apnea and a higher risk of AF. "These data provide compelling evidence that sleep quality itself, even independent of sleep apnea, is an important determinant of AF risk," noted Dr. Marcus. "While there are several available treatments

for AF, prevention of the disease would be ideal. The good news is that sleep quality can be modifiable, and is something that at least to some degree is under the control of the individual. It's possible that improving sleep hygiene, such as performing regular exercise, getting to bed at a reasonable hour on a regular basis, and avoiding viewing screens before bed, as well as caffeine later in the day, might help stave off AF."

Matt Christensen was funded by the Sarnoff Cardiovascular Research Foundation. The Health eHeart Study is supported by the Patient Centered Outcomes Research Network and NIH. The Cardiovascular Health Study is supported by the NIH. The Healthcare Cost and Utilization Project is supported by AHRQ.

HeartRhythm, the official Journal of the Heart Rhythm Society and the Cardiac Electrophysiology Society. For more information: www.heartrhythmjournal.com

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How Do You Assess Pain in Children Who Can't Express Themselves?

Newswise — Pain is a frequent problem for children with complex medical conditions – but many of them are unable to communicate their pain verbally. For these children, nurses face a challenging task in assessing and determining the cause of pain, according to a study in the August issue of the *American Journal of Nursing*. The journal is published in the Lippincott Portfolio by Wolters Kluwer.

"While most kids can be in and out of the primary care provider's office in under an hour for an ear infection, children with complex needs stay in the hospital for a week just to find the ear infection," comments lead author Brenna L. Quinn, PhD, RN, NCSN, CNE, of the Solomont School of Nursing, University of Massachusetts Lowell. "Identifying pain early so teams may get to work finding and addressing the source is essential in avoiding long hospital stays, family stress, poking and prodding, or even surgery and death."

Brenna Quinn added, "When pain assessment Is inadequate, these children suffer unnecessarily."

The two-year study examined the symptoms, diagnostic testing, and nursing assessment of pain in children who have profound intellectual and developmental disability, and are completely dependent on caregivers or medical devices (such as feeding tubes) for their care.

The study included 46 patients seen at a children's hospital Complex Care Service with a chief concern of pain. The patients ranged from infants to young adults; the average age was 13 years. Most had several chronic conditions, most commonly seizure disorders and cerebral palsy; all were unable to verbally communicate their pain – where they were hurting, how much pain they were experiencing, or whether they were in pain at all.

The most common symptoms prompting parents or caregivers to seek medical care for their child were abdominal pain or distention (bloating), irritability, or other signs of pain. In some cases, the parents said that their child was just "not acting like herself [or himself]." The patients underwent an average of five diagnostic tests, most commonly X-rays; and were evaluated by an average of four specialty services while in the hospital.

The most common diagnoses were infections, including: urinary tract infections (30% of children); constipation (20%); and increased seizure activity related to low levels of antiepileptic medications (13%). After other conditions were ruled out, a diagnosis of chronic pain was made in 22% of patients. Across diagnostic groups, symptoms were similar, including irritability, feeding intolerance, mental status changes, and vomiting.

While in the hospital, the children underwent more than 3,300 pain assessments – an average of seven assessments per patient per day. Since the children couldn't communicate their pain, the nurses used a number of assessment tools based on observable pain behaviors (facial expressions, crying, etc.).

Although they represent a small percentage of hospitalizations, medically complex children use a high proportion of healthcare services. "Like all patients, nonverbal children with medical complexity require a balance of standardized and individualized care," Dr. Quinn and coauthors write. Based on their findings, they make recommendations for pain evaluation in this group of patients, including:

- Eliciting the parents' or caregivers' knowledge of the child. As in previous studies, the results suggest that parents are often able to identify behavior changes suggesting that their children are in pain.
- Assessing the presence of pain, even when the child appears to be sleeping or when there are indications of a change in mental status.
- Using pain assessment tools matched to the patient's cognitive abilities, incorporating behavior assessments and input from parents.
- Being alert for potentially life-threatening sources of pain – while also not overlooking more common problems like infections or constipation.
- The researchers emphasize the need for further studies of pain symptoms, evaluation, and diagnosis in children with complex medical conditions, especially those who cannot communicate their pain verbally.

Dr. Quinn and coauthors conclude, "When pain assessment is inadequate or lacking, these children suffer unnecessarily." The researchers are working to develop a tool to aid in comprehensive, efficient assessment of common causes of pain in this vulnerable group of patients.

The *American Journal of Nursing*'s mission is to promote excellence in nursing and health care through the dissemination of evidence-based, peer-reviewed clinical information and original research, discussion of relevant and controversial professional issues, adherence to the standards of journalistic integrity and excellence, and promotion of nursing perspectives to the health care community and the public.

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Inhaled Nitric Oxide May Reduce Kidney Complications from Heart Surgery

The study, "Nitric Oxide Decreases Acute Kidney Injury and Stage 3 Chronic Kidney Disease after Cardiac Surgery," was published online in the American Thoracic Society's American *Journal of Respiratory and Critical Care Medicine.*

Researchers studied 244 adults in Xi'an, China, who underwent surgery to replace more than one heart valve. Because of the duration of the procedure, the patients required placement on cardiopulmonary bypass (a heart-lung machine) for at least 90 minutes.

"Previous studies showed that prolonged cardiopulmonary bypass causes disruption of circulating red blood cells and the release of hemoglobin, which can cause acute kidney injury, leading to kidney failure, and the need for long-term hemodialysis," said lead study author, Lorenzo Berra, MD, Medical Director of Respiratory Care at Massachusetts General Hospital in Boston and Assistant Professor at Harvard Medical School. "We tested whether administration of nitric oxide, a gas normally produced by cells in the lining of blood vessels, might render hemoglobin 'inert,' thereby, decreasing the risk of both acute and chronic kidney injury."

The authors found that patients who received 80 parts per million of nitric oxide (NO), during and for 24 hours after surgery, were less likely to develop acute kidney injury, with a decrease from 64% in the placebo-treated patients to 50% in those who received NO.

The risk of progressing to more serious kidney disease (Stage 3 Chronic Kidney Disease) was also reduced at 90 days, with a decrease from 33% in the placebo-treated patients to 21% in those who received nitric oxide. After one year, 31% in the placebo group had serious kidney disease compared to 18% in the nitric oxide group.

There was also a decrease in the overall mortality rate after one year, from 6% in the placebo group to 3% in the nitric oxide group. This decrease did not reach statistical significance, possibly because of the relatively small number of patients included in the study, the researchers wrote.

According to the authors, several drugs have been tested and shown to be ineffective at protecting the kidneys after cardiac surgery. This is the first study to show that a pharmacological treatment can reduce acute and chronic kidney injury resulting from cardiac surgery.

Importantly, the authors noted that administration of nitric oxide gas appears to be safe: NO delivery did not have to be reduced or stopped in any of the patients who received the gas.

The authors caution that study results may not be generalizable to all cardiopulmonary bypass patients. In the Chinese study, all patients underwent the same type of surgery, and most of the patients were young (average age: 48) because their heart valve problems were caused by rheumatic fever. In North America and Europe, degenerative heart disease is a more common cause of valve dysfunction, and these older patients are more likely to have additional medical problems.

The researchers are now conducting a similar trial at the Massachusetts General Hospital to determine whether nitric oxide provides similar benefits as those seen in the Chinese study.

Compared to the younger, relatively healthy patients in the Chinese study, Dr. Berra said, "We believe that the older patients with an increased number of cardiovascular risk factors, including obesity, hypertension and diabetes, may derive even greater benefit from nitric oxide administration during and after heart surgery."

This study was funded by the National Natural Science Foundation of China, the Xijing Hospital Foundation, the National Key Technology Research and Development Program at the Ministry of Science and Technology of China, the Changjiang Scholars and Innovative Research Team at the University of China, and the Department of Anesthesia, Critical Care and Pain Medicine at Massachusetts General Hospital.

The AJRCCM (The American Journal of Respiratory and Critical Care Medicine) is published by the American Thoracic Society. The American Thoracic Society (ATS) is dedicated to advancing pulmonary, critical care and sleep medicine. It publishes three journals: the American Journal of Respiratory and Critical Care Medicine, the American Journal of Respiratory Cell and Molecular Biology and the Annals of the American Thoracic Society.

UH Rainbow Babies & Children's implants Transcatheter Heart Valve without Surgery in One of the Smallest Patients Ever Recorded

A two-year-old boy with a Complex Congenital Heart Disease underwent successful Transcatheter Pulmonary Valve replacement procedure at University Hospitals Rainbow Babies & Children's Hospital (UH Rainbow). Although this technology has been around for many years, this patient weighing just over 10 kilograms (23 pounds) is one of the smallest patients on record to receive the transcatheter valve without surgical assistance. The patient received the Melody Transcatheter Pulmonary Valve and was discharged two days later.

"Patients with severe forms of Congenital Heart Dsease often require multiple open-heart surgeries during their lives. Having to go through these surgeries is very hard, especially for infants and young children, and this can be particularly stressful to their parents and families," says Martin Bocks, MD, Director, Pediatric Interventional Cardiology at UH Rainbow. "This patient had already undergone multiple heart catheterization procedures and a heart surgery, so our goal was to replace the valve without putting the patient through another operation, and we are thrilled with the results."

The Melody™ Transcatheter Pulmonary Valve (TPV), from Medtronic, was the first medical device of its kind to be approved by the FDA for the treatment of patients with this form of heart disease. The procedure allows the interventional cardiologist to deliver a replacement valve through a catheter requiring only a small incision, in this case through the vein in the patient's neck. The Melody valve has now been implanted in more than 13,000 patients worldwide.

Dr. Bocks, who came to UH Rainbow from the University of Michigan two years ago, has extensive experience with the Melody TPV and has been implanting the device since it was first FDA approved in

2010, though none quite as challenging as his most recent patient.

"Smaller patients have smaller blood vessels and less room to move large catheters, and this anatomic challenge often eliminates the transcatheter route for valve replacement as an option," says Dr. Bocks. "In my field of pediatric interventional cardiology, we continually push the envelope of what we can do in a less or minimally invasive manner. We are all looking for ways to limit pain and discomfort while getting the patients the best possible end result."

Dr. Darren Berman, Co-Director of Cardiac Catheterization and Interventional Therapy in the Heart Center at Nationwide Children's Hospital in Columbus, also has extensive experience with transcatheter valve therapies in patients with complex congenital heart disease. Dr. Berman assisted Dr. Bocks with the transcatheter valve procedure in this patient as part of The Congenital Heart Collaborative. Dr. Berman has researched the topic of transcatheter valve replacement in small children and published the first report on its use in patients less than 30kg.

"In our paper we demonstrated that the Melody valve can be safely delivered in smaller patients and this early experience was instrumental in helping Dr. Bocks and I feel confident that we would be successful in placing a valve even in a patient this small," says Dr. Berman. "This case highlights the invaluable importance of collaboration amongst colleagues and how the institutional collaboration and formation of The Congenital Heart Collaborative benefited this patient," commented Dr. Berman.

The Congenital Heart Collaborative, formalized three years ago, is a partnership between UH Rainbow in Cleveland and Nationwide Children's in Columbus, which brings together expert physicians, surgeons and teams to provide world class care for patients and families in Northeast Ohio. The Congenital Heart Collaborative provides expert heart care for patients from infancy to adulthood. For more information about heart care at UH Rainbow visit rainbow.org/heart.

UH Rainbow Babies & Children's offers the most expansive pediatric care network in Northeast Ohio, with more than 740,000 annual patient encounters at 131 service locations. The vast network includes nationally-ranked UH Rainbow Babies & Children's Hospital -- Cleveland's only full-service, freestanding children's hospital with a medical staff of more than 745 physicians and Northern Ohio's only Level I Pediatric Trauma Center – along with a primary care network of more than 200 pediatric and family medicine providers at 83 offices, inpatient care, pediatric emergency services, urgent care centers, surgery centers, medical and surgical specialty clinics, and advanced newborn and maternal/fetal medicine services. Learn more at Rainbow.org.

Founded in 1866, University Hospitals serves the needs of patients through an integrated network of 18 hospitals, more than 50 health centers and outpatient facilities and 200 physician offices in 15 counties throughout northern Ohio. The system's flagship academic medical center, University Hospitals Cleveland Medical Center, located on a 35-acre campus in Cleveland's University Circle, is affiliated with Case Western Reserve University School of Medicine. The main campus also includes University Hospitals Rainbow Babies & Children's Hospital, ranked among the top children's hospitals in the nation; University Hospitals MacDonald Women's Hospital, Ohio's only hospital for women; and University Hospitals Seidman Cancer Center, part of the NCI-designated Case Comprehensive Cancer Center. UH is home to some of the most prestigious clinical and research programs in the nation, including cancer, pediatrics, women's health, orthopedics, radiology, neuroscience, cardiology and cardiovascular surgery, digestive health, transplantation and urology. UH Cleveland Medical Center is perennially among the highest performers in national ranking surveys, including "America's Best Hospitals" from *U.S. News & World Report.* UH is also home to Harrington Discovery Institute at University Hospitals – part of The Harrington Project for Discovery & Development. UH is one of the largest employers in Northeast Ohio with 26,000 employees.

UH's vision is "Advancing the science of health and the art of compassion," and its mission: "To Heal. To Teach. To Discover." Follow UH on Facebook @UniversityHospitals and Twitter @UHhospitals. For more information, go to UHhospitals.org.

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Pediatric Heart Failure / Heart Transplant Cardiologist
Rank Dependent on Qualifications / Clinical or Tenure Track
Division of Pediatric Cardiology

The Division of Pediatric Cardiology at the University of Utah School of Medicine has an immediate opening for a Pediatric Cardiologist with expertise in heart failure/heart transplant. This is an outstanding opportunity to join a vibrant and collegial academic environment and work in a children's hospital ranked as one of the best in the US. Responsibilities will include focused care to inpatients and outpatients with cardiomyopathy, heart failure due to either cardiomyopathy or congenital heart disease, those supported on a ventricular assist device and patients pre- and post-heart transplantation. In addition to clinical service, there is an expectation for academic work, including teaching, research, administration, as well as advocacy. There will be protected time for clinical research with mentoring available within the Division. Depending on experience, there may be opportunities for leadership responsibilities.

The Heart Transplant Program performs an average of 8-12 heart transplants per year and provides care for over 100 post-transplant patients. Activity in the Mechanical Circulatory Support Program will include patient selection for ventricular assist device support and follow up care in conjunction with cardiothoracic surgeons. Clinical activities will be carried out at Primary Children Hospital and the Division of Pediatric Cardiology affiliated outreach sites.

Qualified candidates must have an M.D. or D.O. degree, be Board Qualified/Board Certified in Pediatric Cardiology, and must have advanced training or expertise in heart failure/heart transplant and mechanical circulatory support. The selected candidate will receive a faculty appointment in the Department of Pediatrics on the Clinical or Tenure track at the academic level commensurate with experience and qualifications.

The University offers a competitive salary and an unmatched benefits program, including non-contributory retirement contributions of 20.2% of annual salary that vest immediately. The Department offers an education loan repayment program, in addition to a faculty development and mentoring program designed to help faculty succeed in translational or basic research.

Salt Lake City offers an incredible quality of life with a growing economy, rich cultural scene with ballet, theatre, symphony, opera and museums, outstanding restaurants, and a moderate cost of living. The city is a ski destination and a gateway to the state's renowned landscapes. In addition to its 14 ski resorts, Utah boasts five national parks (with five more within a day's drive), a variety of golf courses allowing for year-round play, hundreds of miles of hiking and biking trails, and numerous other outdoor activities.

Interested individuals can apply for the position at:
http://utah.peopleadmin.com/postings/82504.
Cover letter and curriculum vitae will be required.

For additional information about the position, please contact:
Lloyd Y. Tani, M.D., Division Chief,
at Lloyd.tani@hsc.utah.edu.

The University of Utah Health Sciences Center is a patient focused center distinguished by collaboration, excellence, leadership, and Respect. The University of Utah HSC values candidates who are committed to fostering and furthering the culture of compassion, collaboration, innovation, accountability, diversity, integrity, quality, and trust that is integral to the mission of the University of Utah Health Sciences Center.

The University of Utah is an Affirmative Action/Equal Opportunity employer and does not discriminate based upon race, national origin, color, religion, sex, age, sexual orientation, gender identity/expression, status as a person with a disability, genetic information, or Protected Veteran status. Individuals from historically underrepresented groups, such as minorities, women, qualified persons with disabilities and protected veterans are encouraged to apply. Veterans' preference is extended to qualified applicants, upon request and consistent with University policy and Utah state law. Upon request, reasonable accommodations in the application process will be provided to individuals with disabilities. To inquire about the University's nondiscrimination or affirmative action policies or to request disability accommodation, please contact: Director, Office of Equal Opportunity and Affirmative Action, 201 S. Presidents Circle, Rm 135, (801) 581-8365.