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## **Case Study of Severe Hemolysis Following Atrial Septal Defect Closure**

By Pooja Nawathe, MD; Myriam Almeida-Jones, MD; Christina Wheelwright, NP; Evan M. Zahn, MD

Indexing Words: Transseptal catheterization, Closure, ASD/PDA/PFO, Complications, Pediatric Catheterization/ Intervention.

#### Introduction

Transcatheter closure of Secundum Atrial Septal Defects (ASD) is well-documented to be an effective alternative to surgical ASD closure.1 The overall complication rate of transcatheter ASD occlusion has been found to be significantly less than that of surgery.<sup>2</sup> There have been several reviews of transcatheter ASD closure devices<sup>2-4</sup> discussing rare, but major adverse events, such as device embolization, cardiac perforation, and thromboembolic complications. We report a case of severe hemolytic anemia after transcatheter ASD closure possibly related to the interface between the device surface and a previously present high velocity jet of mitral regurgitation. The hemolysis resolved quickly following replacement of the initial ASD occluder with a device with different surface characteristics.

#### Case

A 2-year-old male with Trisomy 21 and Rastelli Type A complete atrioventricular (AV) canal defect with well-balanced atrioventricular valves and mildly hypoplastic right ventricle underwent pulmonary artery banding and Patent Ductus Arteriosus (PDA) ligation at three months of age. This was followed by complete surgical repair at nine months of age using a two-patch technique to close his Atrial and Ventricular Septal Defects (ASD,VSD), suture repair of bilateral atrioventricular valves, and debanding the pulmonary artery. Based upon pre-operative concerns of bidirectional shunting at ventricular level and mild right-ventricle hypoplasia, a 6 mm atrial level communication was created at the time of surgery. The post-operative course was uncomplicated. Pre-discharge echocardiogram revealed no residual VSD, tiny residual PDA, trivial right atrioventricular valve regurgitation and moderate left atrioventricular valve regurgitation centrally and through a small residual cleft directed toward the atrial septum.

The patient's growth curve plateaued at one year of age, and failure to thrive persisted despite optimizing caloric intake. An echocardiogram at two years of life showed pure left-to-right atrial level shunting through the ASD, which at that time measured 11 mm, and was associated with right heart dilation, mild tricuspid regurgitation, and unchanged moderate mitral regurgitation (Figure 1). After discussion among our multidisciplinary team, a decision was made to proceed with transcatheter ASD device closure in an attempt to minimize any cardiac contribution to his failure to thrive.

At cardiac catheterization, a small residual PDA was noted and occluded with an Amplatzer Duct

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Occluder (ADO) II (Abbott, Sylmar, CA). Under transesophageal echocardiographic guidance, an 11 mm Amplatzer Septal Occluder (ASO) (Abbott, Sylmar, CA) was implanted uneventfully across the atrial septum. Due to the previous question regarding adequacy of the right heart volume, the patient was monitored for 20 minutes with the device in place prior to release from the delivery cable, so-called, test occlusion. As hemodynamics remained stable, the device was released with complete defect occlusion documented in the catheterization laboratory. Postprocedure transesophageal echocardiogram showed no device impingement upon the atrioventricular valves and unchanged moderate mitral regurgitation. Aspirin was started (5 mg/kg daily) as protocol post septal device placement.

Overnight, the patient developed redtinged urine. Transthoracic echocardiogram confirmed stable device placement, with no residual PDA or ASD shunting. Importantly, there remained moderate mitral regurgitation with a high velocity jet directed towards the inferior aspect of the septal occluder (Figure 2). Laboratory investigations were consistent with an acute hemolytic process: decrease in hematocrit from 37% to 26%, elevated lactate dehydrogenase of 1727 U/L, low serum haptoglobin <6 mg/dl, indirect hyperbilirubinemia of 1.4 mg/dl, and hemoglobinuria. A peripheral blood smear showed normocytic, normochromic anemia with schistocytes. As there were no residual shunts, we hypothesized that the mitral regurgitant jet directed at the inferior aspect of the septal occluder was resulting in contact hemolysis, and adopted a conservative approach. Hydration therapy was initiated to prevent hemoglobinuriarelated renal injury; however, over the ensuing days, the hemoglobin and hematocrit continued to decline requiring multiple transfusions of packed red blood cells on Day 3, 9, and 13 post-procedure (Figure 3). Pentoxifylline was started on Day 9 with no obvious improvement.

On Day 13 the child returned to the catheterization laboratory where the device was removed using a 10 mm Amplatz Goose Neck snare (Medtronic, Minneapolis, MN). A 25 mm Gore Cardioform Septal Occluder (W.L. Gore & Associates) was implanted uneventfully. Post-procedure echocardiograhy confirmed stable device position, no residual interatrial shunting, and unchanged moderate mitral regurgitation still directed medially toward the inferior aspect of the septal occluder (Figure 4). The patient's urine color normalized within six hours of the procedure, hemoglobin and hematocrit remained stable, and lactate dehydrogenase began to normalize. The patient was discharged three days after atrial septal occluder exchange, with no



Figure 1. Pre-procedural transesophageal echocardiogram. Modified 4-chamber views of 2-dimensional and color Doppler images demonstrating an 11 mm ASD (dotted line) and a medial jet of mitral regurgitation (arrow) emanating from a surgically repaired cleft mitral valve (asterisks) and directed toward the inferior aspect of the defect. LA = left atrium. RA = right atrium.



Figure 2. Post-procedural transesophageal echocardiogram. Modified 4-chamber views of 2dimensional and color Doppler images demonstrating color turbulence from a medial jet of mitral valve regurgitation (solid arrow) as it interacts with the inferior portion of the ASO (broken arrow). Note the color turbulence at the left atrial disc as a result of the mitral regurgitation. ASO = Amplatzer Septal Occluder.

further evidence of hemolysis on continued clinical follow-up.

#### Discussion

Documented cases of hemolysis have occurred after transcatheter device closure of



Figure 3. Hematocrit trend related to post procedure days. Day 0 is the day the ASO was implanted. Arrows represent blood transfusions. ASO was removed on Day 13 and replaced with a Cardioform Septal Occluder (asterisk). RBC = red blood cells.

There have also been rare cases of hemolytic anemia reported following surgical patch repair of ASD in the setting of primum ASD repair. In both cases reported by Alehan et al<sup>9</sup> there was associated moderate mitral regurgitation, and the authors hypothesized that the collision of the regurgitant mitral jet with the rough-surfaced Teflon patch used for the repair resulted in the hemolysis. These patients were treated with reoperation and further correction of their mitral regurgitation with the authors concluding that, "foreign materials in association with localized intracardiac turbulence may cause severe hemolysis".<sup>9</sup>

To our knowledge, there have been no similar reports of atrial septal occluders contributing to hemolysis in the presence of



Figure.4. Post-procedural transesophageal echocardiogram after Cardioform Septal Occluder placement. Modified 4-chamber views of 2-dimensional and color Doppler images. The medial jet of mitral regurgitation (solid arrow) continues to be directed at the inferior margin of the device (broken arrow), although there appears to be less turbulent color flow between the discs as compared with imaging after ASO placement (Figure 2).

PDA and VSD, related to residual flow through and around a device, presumably secondary to shear stress placed on red blood cells as they pass at high velocity through and around device matrices.<sup>5-7</sup> Postprocedural hemolysis has been reported only once before in the setting of device closure of ASD[8]. In that case report, Lambert et al suggested that the trajectory of tricuspid regurgitation toward the ASD occluder (Amplatzer 24 mm) was directed through a second unrepaired ASD, and was responsible for the hemolysis. The patient was successfully treated with Amplatzer device removal and surgical repair of both ASDs.

pre-existing mitral regurgitation, which resolved after transcatheter device replacement. The ASO is a remarkably versatile device that has been used successfully in thousands of patients with ASD over the years. While reported major adverse events remain low, the device is known to have a somewhat rough textured surface due to its wire mesh construction. It is this abrasive surface which may in fact contribute to the rare reported cases of erosion.<sup>10</sup> In the case reported herein, we believe there was strong color Doppler echocardiographic evidence that the high velocity medial jet of mitral regurgitation

interacted with the rough surface of the inferior margin of the device, thereby resulting in hemolysis (Figure 2). This is supported by the timeline, the relationship of ASO device implantation and removal with resolution of hemolysis. That being said, numerous patients have undergone successful ASD closure with ASO in the setting of mitral regurgitation, and this should not be considered a contraindication to implanting this device. However, we believe it is likely that in this unusual case, the smoother surface of the Cardioform device, while still interacting with the same high velocity jet of mitral regurgitation, did not result in hemolysis, as there were less abrasive forces imposed upon the red blood cells.

#### Conclusion

Hemolysis should be considered a rare, but possible side effect related to Atrial Septal Defect occlusion with ASO, in the setting of atrioventricular valve regurgitation directed toward the atrial septum. In such a case, consideration should be given to using an alternative device with a smoother surface.

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## **Coronary Artery Fistulas Entering the Right Atrium – A Case Series**

By Riki Patel, BA; Donald Mattia, BS; Randy Richardson, MD

#### Keywords

Coronary Artery Fistula, Right Atrium, Percutaneous Closure, Median sternotomy

#### Introduction

Coronary Artery Fistulas (CAF) are rare cardiac defects that occur from aberrations of normal embryological development. In normal development endothelial outgrowths penetrate the myocardium to form trabecular spaces called sinusoids, which allow for communication with heart cavities. As the heart develops these sinusoids are replaced with a complex reticulation of capillaries, arteries, and veins which can all connect with other vessels and chambers. It is persistence of these connections that may lead to coronary artery fistulas.<sup>1</sup>

CAFs are defined as an abnormal connection between a coronary artery and a cardiac chamber or major vessel.<sup>2</sup> The resultant physiological impact is widely differing depending on the origin as well as termination sites. The major sites of origin are the right coronary artery (RCA) (56%) and the left coronary artery (LCA) (35%); while the major termination sites are the right ventricle (40%), right atrium (26%), and the pulmonary arteries (17%).3 Most CAFs are small and asymptomatic, and thus non-disrupting to myocardial perfusion. However, significant fistula can lead to hemodynamic compromise manifesting as various complications; including myocardial ischemia, heart failure, infarction, endocarditis, rhythm abnormalities, thrombosis of fistula and aneurysms.4 Whether complications are present or not, patients often present with a murmur which prompts further evaluation, and diagnosis is determined through echocardiography and CT angiography. Treatment may not be required if the fistula is clinically silent. However, some fistulas do require surgical or catherization closure - examples include small coronary fistulas that tend to grow with age and become symptomatic, or some asymptomatic fistulas with large volumes of blood shunting.5

#### Case 1

A baby girl was born at term to a G4P4 mother after limited prenatal care. Numerous malformations were noted at birth, including an extra digit on the right foot at the heel, micrognathia, cleft palate, and multiple pale skin nodules across her trunk, legs, and back. Radiographs later Conservative management was elected going forward with regular cardiology follow-up. No cardiac symptoms have developed and there has been no growth



Figure Ia (left). A cardiac CTA 3D reconstruction showing a fistulous connection from the right coronary artery (arrow) to the right atrium (blue).

Figure Ib (right). A cardiac CTA 3D reconstruction showing just the fistulous vessels.

revealed 2 tibias and 1 fibula in the right leg. These midline defects prompted further internal organ evaluation, including her heart. On physical exam, a grade 4/6 systolic murmur was heard over the entire chest wall. An EKG showed S-T depression and inverted T waves in the lateral precordial leads, suggestive of subendothelial injury. Further testing with echocardiology showed a Patent Foramen Ovale (PFO), high right ventricular systolic pressure with mild dilation, and severely dilated right and left coronary arteries. Multiple coronary artery fistulas and collaterals were found, mainly from the RCA, that fed a homogenous tissue mass (11x16 mm) appearing to be nearly contiguous with the superior aspect of the atrial septum. It was positioned between the posterior aortic root and SVC, with no evidence of obstruction to either the SVC or right upper pulmonary vein. The mass was echobright and highly vascularized, possibly representing an infantile intracardiac hemangioma. Fistulous connections between the LAD and right ventricle were noted as well. The baby girl was monitored in the NICU after birth for failure to thrive, but was otherwise asymptomatic. After prognosis was discussed, a gastric-tube was inserted for feeding, while all other procedures were ultimately declined by the patient's parents.

or change in the fistulous connection after two years.

#### Case 2

A baby girl at 3 Days of Life was found to have a 3/6 continuous murmur heard over the left sternal border. She underwent echocardiography, which showed an Atrial Septal Defect (ASD) and left coronary artery dilation at the origin, suggesting the presence of a coronary artery fistula. She subsequently underwent an MRI study, which again indicated the presence of a fistula from the left coronary artery going to the right atrium, an ASD, and presence of a QP to QS ratio of 1.7:1. Clinically the child was asymptomatic, eating well, and had normal growth. Conservative management with close follow-up was elected at that point in time. At one year of age, CT evaluation showed significant enlargement of the coronary fistula. The patient was, therefore, brought to the catheterization lab for further delineation and possible occlusion. Angiogram showed severe dilation of the left main coronary artery with the fistula appearing to arise from the circumflex artery, course posteriorly, and insert into the right atrium. Due to stasis of the left main coronary artery with test occlusion, it was thought that a coil or device occlusion of this fistula might



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cardiac catheterization, any patient where the margins of the defect are less than 5 mm to the coronary sinus, inferior vena cava rim, AV valves, or right upper lobe pulmonary vein. The AMPLATZER™ Cribriform Occluder is a percutaneous, transcatheter, atrial septal defect closure device intended for the closure of multifenestrated (cribriform) atrial septal defects (ASD). Patients indicated for ASD closure have echocardiographic evidence of fenestrated ostium secundum atrial septal defect and clinical evidence of right ventricular overload (such as, 1.5:1 degree of left-to-right shunt or right ventricular enlargement). Contraindications: Treatment of patients with PFO defects, this device has not been studied in patients with PFO defects; patients known to have extensive congenital cardiac anomaly, which can only be adequately repaired by way of cardiac surgery, patients known to have sepsis within 1 month piror to implantation, or any systemic infection that cannot be successfully treated prior to device placement, patients known to have a bleeding disorder, untreated ulcer, or any other contraindications to aspiring therapy unless another antiplatelet agent can be administered for 8 months, patients known to have demonstrated intracardiac thrombi on echocardiography (especially left atrial or left atrial appendage thrombi), patients whose size would cause the patient to be a poor candidate for cardiac catheterization, any patient where the radius of the device is greater than the distance from the central defect to the aortic root or superior vena cava

#### AMPLATZER™ VSD OCCLUDERS

The AMPLATZER™ Muscular VSD Occluder is indicated for use in patients with a complex ventricular septal defect (VSD) of significant sizeto warrant closure (large volume left-toright shunt, pulmonary hypertension, and/or clinical symptoms of congestive heart failure) who are considered to be at high risk for standard transatrial or transarterial surgical closure based on anatomical conditions and/or based on overall medical condition. Highrisk anatomical factors for transatrial or transarte¬rial surgical closure include patients requiring left ventriculotomy or an extensive right ventriculotomy, with a failed previous VSD closure, with multiple apical and/or anterior muscular VSDs ("Swiss cheese septum"), with posterior apical VSDs covered by trabeculae. Contraindications: The AMPLATZER™ Muscular VSD Occluder is contraindicated for the following: Patients with defects less than 4 mm distance from the semilunar (aortic and pulmonary) and atrioventricular valves (mitral and tricuspid), patients with severely increased pulmonary vascularresistance above 7 Wood units and a right-to-left shunt and documented irreversible pulmonary vascular disease, patients with perimembranous (close to the aortic valve) VSD, patients with postinfarction VSD, patients who weigh less than 5.2 kg. (Patients smaller than 5.2 kg were studied in the clinical trial, but due to poor outcome, these patients have been contraindicated for device placement. Data from these patients has not been included in the overall analysis.), patients with sepsis (local/generalized), patients with active bacterial infections, patients with contraindications to antiplatelet therapy or agents. The AMPLATZER™ Post-infarct Muscular VSD Occluder is a percutaneous transcatheter occlusion device intended for closure of post-myocardial infarct muscular VSDs in patients who are not satisfactory surgical candidates. Contraindications: The AMPLATZER™ Postinfarct Muscular VSD Occluder is contraindicated for the following: patients with perimembranous VSDs or a VSDs close to the aortic or mitral valve, patients with congenital muscular VSDs, patients with the presence of thrombus at the intended site of implant, or documented evidence of venous thrombus in the vessels through which access to the defect is gained, patients with active endocarditis or other infections producing bacteremia, patients whose vasculature, through which access to the defect is gained, is inadequate to accommodate the appropriate.sheath size, patients known to have active sepsis or any systemic infection that cannot be successfully treated prior to device placement, any patient known to have a bleeding disorder, untreated ulcer, or any other contraindications to aspirin therapy, unless another antiplatelet agent can be administered for 6 months.

#### AMPLATZER™ DUCT OCCLUDERS

The AMPLATZER<sup>™</sup> Duct Occluder is a percutaneous, transcatheter occlusion device intended for the nonsurgical closure of a patent ductus arteriosus (PDA). **Contraindications:** patients weighing less than 6 kg, patients less than 6 months of age, presence of thrombus at the intended site of implant, or documented evidence of venous thrombus in the vessels through which access to the defect is gained, active endocarditis or other infections producing bacteremia, patients whose vasculature, through which access to the defect is gained, active endocarditis or other infections producing bacteremia, patients whose vasculature, through which access to the defect is gained, is inadequate to accommodate the appropriate sheath size, patients with pulmonary hypertension with pulmonary vascular resistance of greater than 8 Wood units or Rp/ Rs of greater than 0.4

The AMPLATER<sup>™</sup> Duct Occluder II is a percutaneous transcatheter occlusion intended for the nonsurgical closure of patent ductus arteriosus. Contraindications: The AMPLATZER<sup>™</sup> Duct Occluder II is contraindicated for the following: patients weighing less than 6 kg, patients less than 6 months of age, patients with a window-type patent ductus arteriosus (ie, length less than 3mm), patients with an active infection, patients with thrombus at the intended site of implant, patients with pulmonary hypertension with pulmonary vascular resistance of greater than 3 Wood units or Rp/Rs of greater than 0.4, patients with patent ductus arteriosus greater than 12 mm in length by angiography, patients with patent ductus arteriosus greater than 5.5 mm in diameter by angiography.

**CAUTION:** These products are intended for use by or under the direction of a physician. Prior to use, reference the Instructions for Use inside the product carton (when available) or at manuals. sjm. com for more detailed information on Indications, Contraindications, Warnings, Precautions and Adverse Events.

#### Abbott Structural Heart

3200 Lakeside Dr., Santa Clara, CA. 95054 USA, Tel: 1.800.227.9902 Unless otherwise specified, all product and service names are trademarks owned by or licensed to Abbott, its subsidiaries or affiliates. No use of any Abbott trademark, trade name, or trade dress may be made without the prior written authorization of Abbott, except to identify the product or services of the company. ©2018 Abbott. All rights reserved. SJM-AMPLP-0718-0103 | Item approved for US use only. propagate a clot into the left circumflex and LAD. Test occlusion of the fistula under full anticoagulation also resulted in better systolic blood pressure and had no ST segment changes after 5 minutes. More precise closure and ligation was performed via a median sternotomy with vessel clipping. Postoperative course was uneventful and subsequent testing showed resolution of the murmur along with normal oxygen saturations and pulses in all extremities.



Figure II. A cardiac CTA 3D reconstruction demonstrating a dilated left coronary artery (tan) with a fistulous connection to the right atrium (blue).

#### Case 3

A 2.5-month-old baby girl with a past medical history of a muscular Ventricular Septal Defect (VSD) was found to have a holosystolic murmur on routine physical exam. Transesophageal echocardiography (TEE) revealed a single coronary artery. This consisted of a very large, dilated leftcoronary system with a fistula from the left coronary artery into the right atrium and Right Ventricular Outflow Tract (RVOT), which prompted admission to the Pediatric Cardiology ward. Cardiac catheterization again indicated a complex coronary fistula arising off the left-anterior descending artery (LAD), draining through a complex network of collaterals into the right atrium. The tortuous and serpentiginois collateral network was also seen crossing the anterior surface of the right ventricle and eventually draining underneath the pulmonary valve. No right coronary artery was visualized coming off the coronary cusp. Instead, the left main coronary artery was grossly dilated and supplied the entire right coronary system via collaterals. CT angiography on the following day confirmed these findings.

Due to the complexity of the fistula, intraoperative intervention was planned via median sternotomy for the ligation of the fistulous connection. In the operating room, confirm the ligation. The angiogram confirmed appropriate filling of the left coronary system with collateralization with the right coronary system. No further fistulous connections to the right atrium or RVOT were seen. In addition, the complex collateral vessels over the anterior surface of the right ventricle were significantly diminished in size and filling.

The patient was returned to the pediatric ward in stable condition. Two days postoperation, she had elevated troponins as well as ST segment depression, which resolved with nitroglycerin. The patient was extubated and had oxygen saturations greater than 92% in all extremities, which prompted discharge in stable condition at five days post-operation.



Figure IIIa (left). A cardiac CTA 3D reconstruction showing a dilated left anterior descending coronary artery with a large network of fistulous connections draining into the right side of the heart.

Figure IIIb (right). An axial Cardiac CT showing the fistulous vessels specifically entering the right atrium.

gross inspection revealed a large network of dilated vessels on the entire anterior surface of the right ventricle. Using intraoperative angiography, the main fistulous connection diving into the right atrium was able to be visualized. The team then dissected out and ligated the fistula. Directly after this a TEE showed normal systolic function and no residual fistulous connection. At this point, the operative team elected to perform another intraoperative angiography to further

#### Case 4

A 6-year-old boy presented to the ER for fever and leg pain with no significant past medical history. The patient had ongoing symptoms for six days and blood cultures drawn in the ER were later found to be positive for S. Aureus. Antibiotic coverage with cefazolin was started and the patient was admitted for bacteremia and further evaluation. An MRI done at this time showed soft tissue and muscular infection of the left thigh.



## Archiving Working Group

International Society for Nomenclature of Paediatric and Congenital Heart Disease ipccc-awg.net During his admission, a systolic murmur was h e a r d o n p h y s i c a l e x a m a n d a n echocardiogram was performed in an attempt to determine the source of bacteremia. The results showed significant dilation of right-coronary artery measuring up to 9.3mm. The echocardiogram also revealed a 16mm x

17mm mass in the right atrium, which at the time was believed to be a thrombus.

A cardiac CT was then done to further delineate the nature of the dilation, as well as the mass. It demonstrated that the patient had a right coronary artery fistula, measuring



Figure IVa (left). A cardiac CTA 3D reconstruction showing a fistulous connection from the right coronary artery (arrow) to the right atrium (blue).

Figure IVb (right). A cardiac CTA 3D reconstruction showing just the fistulous vessel course.



Figure IVc. An axial cardiac CT showing a coronary artery fistula entering the right atrium (arrow).

around 21mm with the posterior floor of the right atrium. After this finding, a subsequent echo was done which showed that the right coronary fistula had an aneurysmal opening with turbulent flow at the site of entry into the right atrial floor. Based on these findings, the cardiac team decided that the fistula could be ligated in the catheterization lab without having to do a sternotomy. A flow jet from the fistulous communication was also discovered, creating a rough surface which was serving as a nidus for bacterial adhesion and resulting in MSSA endocarditis.

The patient was taken to the catheter lab and had the fistula closed with a 8mm vascular plug. The procedure was tolerated well and no complications were initially seen. Anticoagulation was started prophylactically, as there was some concern for possible thrombus formation in the RCA due to sluggish flow. The patient was discharged and in stable condition. Seven days later though, the patient was at school and started to feel dizzy and nauseous. He was taken to the ER and an EKG showed first degree heart block with a PR interval of 321 ms, which prompted admission for further evaluation. The next day the patient progressed to Type Three heart block and was taken to the ICU. Cardiology was consulted and they hypothesized that the heart block was due to cardiac remodeling from the vascular plug procedure, which was interfering with the electrophysiology of the heart. Solumedrol was started and the patient converted back to sinus rhythm. A pacemaker was being considered at this point, but as the patient was in sinus rhythm, no further intervention was done and the patient was discharged.

#### Discussion

Coronary artery fistulas represent a rare heart defect, having a 0.002% incidence in the general population with no predilection for either gender.<sup>6,7</sup> There are two main classifications of fistulas. The more common type is congenital, but acquired fistulas can also occur after trauma or surgery. Here we present four clinical cases of congenital coronary artery fistulas coursing to the right atrium. All of these patients were asymptomatic at birth, but several developed complications of the disease that lead to hospital presentation. This is the typical course of coronary artery fistula presentations, as most are small during childhood because myocardial blood flow is not compromised. Symptoms develop later on in life due to the fistulas growing with age, leading to a coronary blood-steal phenomenon in which the myocardium becomes underperfused.<sup>1</sup> Untreated fistulas cause clinical symptoms in only 19% of patients under 20 years old, but that number increases to 63% for patients who are 20 vears or older.8 As expected from myocardial ischemia, the most common symptom reported is dyspnea with exertion. These fistulas also

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commonly cause left-to-right shunts within the heart, leading to a state of volume overload that results in congestive heart failure. Arrhythmias, pulmonary hypertension, premature coronary atherosclerosis, aneurysms, rupture or thrombosis of the fistula, palpitations, hemoptysis, paroxysmal nocturnal dyspnea, edema, endocarditis, valvular regurgitation (secondary to papillary muscle dysfunction), and angina are other less commonly found symptoms.<sup>6</sup>

After age 20, complications may begin to arise due to fistula growth. Symptoms are either caused by a left-to-right shunt or arrhythmias, such as atrial fibrillation and ventricular tachyarrhythmias, due to excessive cardiac chamber load. Infective endocarditis has been reported in up to 12% of patients, which was how Case 4 initially presented. About 20% of patients with coronary artery fistulas have additional cardiac anomalies like Tetralogy of Fallot (TOF), aortic atresia, pulmonary atresia, Atrial and Ventricular Septal Defects, and patent arterial duct.<sup>5,6</sup>

Three of our patients were asymptomatic, aside from Case 4 which presented with fever and muscular infection secondary to infective endocarditis. The common factor between all of our patients' presentations was the presence of a murmur on physical exam. The murmurs presented here were either continuous or systolic murmurs. Literature searches suggest that the timing of the murmur and the location on the chest wall where the murmur is loudest depends on where the fistula inserts into the heart, along with any other structural abnormalities.<sup>9</sup>

After detection of these murmurs, further investigation generally began with an EKG and transthoracic echocardiography, which gave valuable information concerning the blood flow through the layers of the heart and the structure respectively. Next steps include transesophageal echocardiography and cardiac magnetic resonance imaging, which are more invasive, but gave an even clearer anatomical picture along with insight into flow velocity and direction. To confirm the presence of a fistula, coronary angiography remains the gold standard.<sup>9</sup> The major benefit of angiography is that it helps determine which coronary arteries are involved with the fistula, along with communicating chamber/vessel locations. This will give the greatest level of detail, which is needed to evaluate optimal intervention.

There are several options available for treatment of coronary artery fistulas. Three of the presented patients were young and asymptomatic, presenting simply with a murmur. If subsequent testing reveals a coronary artery fistula, it may be appropriate to monitor and educate caretakers of warning signs/symptoms. This was the elected course in two of the three cases. In rare instances (1%-2%), spontaneous closure of a fistula can occur secondary to spontaneous thrombosis.<sup>10</sup> The main indication for closure of a fistula is a significant left-to-right shunt that causes ventricular overload. This shunting is commonly known as coronary steal, where blood from the high-pressure fistula flows into the low-pressure chambers of the heart. This can lead to hypertrophy and ischemia. Early surgical correction is indicated to prevent these instances when a Qp to Qs ratio (pulmonary flow:systemic flow) is greater than 1.5.11 This was the scenario for Case 2, in which the patient was closely followed until this overload condition was met, and subsequently treated surgically.

The options for fistula closure include percutaneous transcatheter occlusion and surgical ligation via median sternotomy. Generally,

catheterization is initiated for potential correction of the fistula. Test occlusion of the fistula can be achieved by balloon catheterization, as demonstrated in Case 2. Ultimately, the treatment chosen is determined by the coronary artery involved, location of the fistula, drainage site of the fistula, and risk of thrombosis or ischemia pending occlusion.<sup>1</sup> Mavroudis et al. determined a criteria for when percutaneous embolization could be used over median sternotomy surgical repair:

- 1. The presence of only one coronary artery fistula
- 2. A narrow fistula drainage site
- 3. Absence of large branch vessels
- 4. Safe and direct accessibility to the artery feeding the fistula.

If any of these four criteria are not met, surgical ligation is the suggested treatment option.<sup>12</sup> Thus, surgical treatment was chosen for Case 3, as the complexity of the fistula would allow for poor percutaneous repair.

Transcatheter approaches provide a greater benefit over median sternotomy by limiting potential iatrogenic injuries. They also are less expensive, have a decreased recovery time, and better cosmetic results.<sup>12</sup> Overall, there is a very low rate of morbidity, mortality, or recanalization when CAF's are closed in infancy or childhood as detection, diagnosis, and treatment have improved over time. Ultimately the benefits of recovery time and fewer traumatic injuries associated with the transcatheter approach need to be weighed against the ability to adapt to complexity for each unique case.

#### Conclusion

Coronary Artery Fistula are rare cardiac defects that are defined as an abnormal connection between a coronary artery and a cardiac chamber or major vessel.<sup>2</sup> CAF's are most commonly asymptomatic, but can cause an array of effects including: myocardial ischemia, endocarditis, and congestive heart failure. Our cases were a great example of how CAF identification is often first suspected through auscultation of a murmur, but can also be from complications of the fistula. This suspicion leads to further testing, in the form of echocardiography, cardiac MRI, coronary CT - with coronary angiography being the gold standard. Symptomatic fistulas will most often be treated with either surgical or catheter closure of the vessel based on Mavroudis criteria, but monitoring is appropriate if no symptoms or shunting are present. Overall, CAFs are rare cardiac defects and a high index of suspicion is needed to assess, diagnose and treat this condition.

#### **Compliance with Ethical Standards**

The authors declare that they have no conflict of interest. This study was not funded. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Because this was a retrospective study, for this type of study, formal consent is not required.

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## **Medical News, Products & Information**

Compiled and Reviewed by Kate Baldwin, Special Projects Editor

## Abbott Receives FDA Approval for Next-Generation MitraClip<sup>®</sup> Device to Treat People with Leaky Heart Valves



After PT-Valve implantation

On July 12<sup>th</sup>, 2018, Abbott announced it received approval from the U.S. Food and Drug Administration (FDA) for a next-generation version of its leading MitraClip<sup>®</sup> heart valve repair device used to repair a leaky mitral valve without open-heart surgery. The transcatheter clip-based therapy, a third generation of product innovation, has been used to treat more than 65,000 patients worldwide over the last ten years.

The next-generation MitraClip system provides cardiologists with advanced steering, navigation, and positioning capabilities for the clip, making it easier to use in difficult anatomies. The enhanced system is designed to allow for more precise placement during deployment, resulting in more predictable procedures, and additionally offers a second clip size with longer arms that expands the reach of the clipbased device. The additional clip size is designed to help doctors treat patients who have more complex anatomies when repairing the mitral valve. Abbott received CE Mark for the next-generation device earlier this year, allowing for sale of the devices in the European Union and other countries that recognize this regulatory designation.

"Physicians rely on MitraClip as an alternative to surgery for patients who aren't surgical candidates and may need treatment to relieve their symptoms or to survive," said Prof. Francesco Maisano, UniversitätsSpital Zürich, Switzerland, who was an early implanter of MitraClip. "The enhanced MitraClip design allows for even more precise navigation, accuracy, and stability during valve repairs, which may be important when treating people with more complex or advanced valve disease."

A leaking mitral valve, known as mitral regurgitation (MR) is a serious, progressive heart disease in which the flaps of the mitral valve do not close properly, allowing blood to flow backward into the heart. Incidence of mitral regurgitation increases with age, with nearly one in 10 people over the age of 75 having moderate to severe disease.<sup>1</sup> Before MitraClip, people who were not eligible for the standard-of-care surgery to treat their MR could only manage their symptoms with medications that don't stop the progression of the disease. Left untreated, MR leads to a variety of life-altering symptoms and severe complications, and may ultimately lead to heart failure and death.<sup>1,2,3</sup>

"Abbott engineers designed these enhancements based on feedback from doctors to improve device delivery and to treat more types of cases and anatomies," said Michael Dale, vice president for Abbott's structural heart business. "We're committed to helping people with mitral regurgitation return to living their best lives, and these advances will enable doctors to treat even more patients without surgery."

MitraClip treats people with degenerative mitral regurgitation and is a therapy that is delivered via a catheter to the heart through a blood vessel in the leg. MR patients are often not eligible for the standard-ofcare surgery because of advanced age, frailty, multiple comorbidities or other complicating factors and the therapy offers a minimally invasive alternative. Treatment with MitraClip provides almost immediate symptom relief and patients are released from the hospital on average after two days.

Abbott recently began enrollment in the MitraClip EXPAND clinical study, a prospective study evaluating the safety and performance of the new MitraClip system in a contemporary real-world setting. Saibal Kar, MD, Director of Interventional Cardiac Research at the Smidt Heart Institute at Cedars-Sinai in Los Angeles, Calif., treated the first patient enrolled, and is the lead investigator of the study. EXPAND will enroll approximately 1,000 patients in more than 50 centers across the U.S. and Europe and interim results from the study are expected later this year.

MitraClip received initial CE Mark approval in Europe in 2008, and was approved by the FDA in 2013. By securing a portion of the leaflets of the mitral valve with an implanted clip, the heart can pump blood more efficiently throughout the body, thereby relieving the symptoms of severe MR and improving patient quality of life.<sup>4</sup>



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- Pediatric Cath Lab experience is preferred but not required.

#### About the Practice:

The Congenital Heart Center, established in 2010, has been ranked as one of the top-50 pediatric heart centers in the country by U.S. News and World Report for the last seven years, in the top 25 for the last two years. Our comprehensive services include cardiac imaging, electrophysiology, dedicated cardiovascular intensive care staff, and regional referral programs in heart failure / transplantation, adult congenital heart disease, and fetal echocardiography. Surgical and cardiac catheterization / electrophysiology volume have been growing at a rate of 15% per year over the last 6 years. A new state of the art two lab cardiac catheterization and electrophysiology suite opened in February 2017. The cardiac catheterization program is active in industry sponsored clinical research. Our new 3-D printing lab will open in the fall of 2018.

LCH and SHVI are both premier referral facilities within the Atrium Health (formerly Carolinas HealthCare System), one of the nation's leading and most innovative healthcare systems. Atrium operates nearly 2,500 system-employed physicians, more than 60,000 employees and more than 7,460 licensed beds across the Carolinas.

Levine Children's Hospital (LCH), a state-of-the-art, 234 bed facility is the largest and most comprehensive children's hospital between Washington, DC and Atlanta, GA. LCH has a robust inpatient service with a 20-bed PICU/CVICU, 85-bed NICU, and an inpatient rehabilitation facility.

Sanger Heart & Vascular Institute (SHVI) is one of the largest cardiac and vascular programs in the Southeast, with more than 50 years of experience in providing world-class, comprehensive acute and chronic cardiovascular services, including the region's only heart transplant center and pediatric heart catheterization and surgical programs.

#### About the Community:

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#### The Washington Adult Congenital Heart Program Earns Accreditation from the Adult Congenital Heart Association

The Washington Adult Congenital Heart Program is one of the First in the United States to Earn ACHA Adult Congenital Heart Disease (ACHD) Accreditation.

In recognition of its expertise in serving adults with Congenital Heart Disease (CHD), structural heart conditions present at birth, the Washington Adult Congenital Heart (WACH) program, part of Children's National Health System, receives accreditation from the Adult Congenital Heart Association (ACHA), a nationwide organization focused on connecting patients, family members, and healthcare providers to form a community of support and a network of experts with knowledge of Congenital Heart Disease.

Individuals with CHD, the most common birth defect diagnosed in one in 100 births, are living longer. There are 1.4 million adults in the U.S. living with one of many different types of congenital heart defects, ranging among simple, moderate, and complex.

"This accreditation lets patients and other specialists know what to expect if they visit our center," says Anitha John, MD, PhD, a Congenital Heart Disease specialist, and the Director of WACH. "While the field of Congenital Heart Disease is small enough to personalize, it's large enough to standardize."

WACH received accreditation by meeting ACHA's criteria, which includes medical services and personnel requirements, and going through a rigorous accreditation process, both of which were developed over a number of years through a collaboration with doctors, physician assistants, nurse practitioners, nurses, and ACHD patients. "There are now more adults than children in the U.S. with CHD," said Mark Roeder, President & CEO of ACHA. "Accreditation will elevate the standard of care and have a positive impact on the futures of those living with this disease. Coordination of care is key, and this accreditation program will make care more streamlined for ACHD patients, improving their quality of life."

"Data drives decisions," notes Dr. John. "We're happy to work with the nation's largest patient advocacy organization to model standards patients and providers will learn to identify and recognize."

The ACHA ACHD Accredited Comprehensive Care Center designation now applies to 18 programs:

- Ahmanson/UCLA Adult Congenital Heart Disease Center (Los Angeles, CA)
- Adult Congenital Heart Program, Stanford University (Palo Alto, CA)
- Colorado Adult & Teen Congenital Heart (CATCH) Program at University of Colorado Hospital and Children's Hospital Colorado (Aurora, CO)
- Washington Adult Congenital Heart (WACH) program, which is part of Children's National Health System (Washington, D.C.)
- Memorial Regional Hospital/Joe DiMaggio Children's Hospital ACHD Center (Hollywood, FL)
- Boston Adult Congenital Heart (BACH) and Pulmonary Hypertension Program (Boston, MA)
- University of Michigan Adult Congenital Heart Program (Ann Arbor, MI)
- Washington University Adult Congenital Heart Disease Program (St. Louis, MO)
- Adult Congenital Heart Disease Program at Children's Hospital & Medical Center and Nebraska Medicine (Omaha, NE)
- NYU Langone Health Adult Congenital Heart Disease Program (New York, NY)
- Cincinnati Children's Adult Congenital Heart Disease Program (CCHMC) (Cincinnati, OH)
- COACH: Columbus Ohio Adult Congenital Heart Disease & Pulmonary Hypertension Program (Columbus, Ohio)
- Medical University of South Carolina Adult Congenital Heart Disease Program (Charleston, SC)
- UT Southwestern and Children's Health Adult Congenital Heart Disease Program (Dallas, TX)
- Texas Children's Hospital Adult Congenital Heart Program (Houston, TX)
- Adult Congenital Heart Disease Program at University of Washington & Seattle Children's Hospital (Seattle, WA)
- Providence Adult and Teen Congenital Heart Program (PATCH) (Spokane, WA)
- Wisconsin Adult Congenital Heart Disease Program (WAtCH) at Children's Hospital of Wisconsin (Milwaukee, WI)

A study published in *Circulation* examined mortality rates among 70,000 patients living with CHD over a 15-year period, from 1990 to 2005, and saw mortality rates fall with referrals to specialized ACHD care centers.

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## PEDIATRIC CARDIOLOGY NP

JOB ID: 053489

#### Department Description: PCS Nurse Practitioners - Pediatric Cardiology

The Division of Pediatric Cardiology provides comprehensive, centerbased care to children with congenital and acquired heart disease. The Pediatric Heart Center is multi-disciplinary with close cooperation between pediatric cardiologists, surgeons, critical care physicians, anesthesiologists, and nurses. We have created a team committed to outstanding patient outcomes for children with complex heart disease in a patient-centered environment.

#### RESPONSIBILITIES

A Pediatric Congenital heart disease (CHD) catheterization/fetal cardiac catheterization Nurse Practitioner (PNP) provides comprehensive management of patients with congenital heart disease (CHD) or families expecting to deliver an infant with a fetal diagnosis of CHD, emphasizing health promotion and disease prevention, as well as the diagnosis and management of acute and chronic diseases associated with CHD, especially those who require a diagnostic/interventional cardiac catheterization, fetal cardiac intervention or a hybrid intervention. Functions under established standardized protocols and works autonomously as well as in collaboration with other health care professionals to diagnose and manage patients' health care needs before, during and after cardiac catheterization. Responsibilities include ordering, conducting and interpreting appropriate diagnostic and laboratory tests; prescription of pharmacologic agents and treatments and nonpharmacologic therapies in a cost efficient manner, writing inpatient and outpatient NP notes and assist interventionist in the cath lab. Acts as a health care resource, interdisciplinary consultant, patient advocate and mentor for the development of other health care colleagues. Demonstrates commitment to a patient and family focused model of care and assists in the analysis of the health care delivery system to provide optimal patient care. PNPs may participate in the design and implementation of research and/or quality improvement studies within the CHD division.

#### QUALIFICATIONS

#### REQUIRED

- At least one year recent experience working as a Pediatric NP
- Previous critical care experience as RN or NP
- Possession of valid California RN and Nurse Practitioner license
- Furnishing license and DEA within 3 months of hire
- Master's degree from an accredited School of Nursing.
- Graduation from a Pediatric NP Program
- National NP Certification
- Current PALS and BLS Certification
- CITI certification within 3 months of hire if conducting clinical research involving human subjects

#### PREFERRED

- At least 3 years of experience working as a Pediatric NP
- One year experience as a NP in a pediatric CHD/interventional service highly desirable

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The University of California, Davis is an Affirmative Action/Equal Opportunity Employer One of the unique features Dr. John and the WACH team provide to patients is the ability to personalize treatment, understand the science and new types of surgery, and take part in continuing medical education forums.

"We're empowering patients to become an active participant and an engaged member of their medical care team," adds Dr. John.

The Washington Adult Congenital Heart (WACH) program was established in 2001, and is part of Children's National Health System, providing continuity of care for adults with Congenital Heart Disease. The program is led by Anitha John, MD, PhD, a Congenital Heart Disease specialist, and provides treatment for both survivors of early surgery and for newly diagnosed adults. For more information about WACH, visit www.ChildrensNational.org/WACH.

The Adult Congenital Heart Association (ACHA) is a national notfor-profit organization dedicated to improving the quality of life and extending the lives of adults with CHD. ACHA serves and supports the more than one million adults with CHD, their families and the medical community—working with them to address the unmet needs of the long-term survivors of congenital heart defects through education, outreach, advocacy, and promotion of ACHD research. For more information about ACHA, visit www.ACHAHeart.org.

The ACHA ACHD Accreditation Program was partially funded by Actelion Pharmaceuticals U.S., Inc. (https://www1.actelion.us) ACHA and Actelion Pharmaceuticals have partnered together since 2007 to support the CHD community.

#### New Cardiac Imaging Technique Shortens Testing Time and Improves Patient Comfort, Potentially Increasing Diagnostic Accuracy for Heart Disease

A team of Cedars-Sinai investigators has developed a new technique for conducting cardiac magnetic resonance imaging (MRI) tests that improves patient comfort, shortens testing time and has the potential to increase diagnostic accuracy and reliability.

The new technique, dubbed MR Multitasking, is detailed in the April 9<sup>th</sup> issue of *Nature Biomedical Engineering* and solves the problem many cardiologists face when using conventional methods to perform MRI scans: how to get a still image when a beating heart and blood flow can blur the picture.

"It is challenging to obtain good cardiac magnetic resonance images because the heart is beating incessantly, and the patient is breathing, so the motion makes the test vulnerable to errors," said Shlomo Melmed, MB, ChB, Executive Vice President of Academic Affairs and the Dean of the Cedars-Sinai medical faculty. "By novel approaches to this longstanding problem, this research team has found a unique solution to improve cardiac care for patients around the world for years to come."

Currently, the standard approach is to ask patients undergoing cardiac MRI to hold their breath while images are being taken—

but then each image must be timed precisely to a specific part of a heartbeat. Unfortunately, this method has proved difficult, unreliable and unsuitable for patients, especially those who have irregular heartbeats or breathing problems.

The investigators' solution: Rather than trying to avoid the motion caused by breathing and heartbeats, the investigators decided to embrace the movement, said Anthony G. Christodoulou, PhD, a research scientist in Cedars-Sinai's Biomedical Imaging Research Institute and the study's first author.

"Our solution is like making a video instead of a still image," Christodoulou said. "MR Multitasking continuously acquires image data and then, when the test is completed, the program separates out the overlapping sources of motion and other changes into multiple time dimensions."

The *Nature Biomedical Engineering* study shows that MR Multitasking was tested in 10 healthy volunteers and 10 cardiac patients, and was found to be accurate, said Debiao Li, PhD, Director of the Biomedical Research Institute and the primary investigator of the study. It also was more comfortable for patients, he said, because patients were no longer required to hold their breath, and the team was able to complete three cardiac MRI tests in as little as 90 seconds—where current standard approaches would have taken much longer.

"Now we collect all the data throughout the entire test and sort it afterwards," Li said. "We get full control after the test, as opposed to trying to control the body's natural movement during imaging."

Incorporating motion and time into the MR Multitasking analysis means that the images have six dimensions, Christodoulou said.

"If a picture is 2-D, then a video is 3-D because it adds the passage of time," Christodoulou said. "Our videos are 6-D because we can play them back four different ways. We can play back cardiac motion, respiratory motion and two different tissue processes that reveal cardiac health."

MR Multitasking is already in use at several medical centers in the U.S. and China, Li said. The team now is looking to expand MR Multitasking to patients with other diseases, like cancer.

"People have to breathe no matter what disease they have, so the ability to separate out the motion is important to many medical specialties," Li said.

The investigators' work was supported by grants from the National Institutes of Health, NIH 1R01HL124649 and NIH T32HL116273. The DOI for the study is dx.doi.org/10.1038/s41551-018-0217-y.

## What Effect Do New Guidelines Have on Prevalence of High Blood Pressure in Children?

*Bottom Line:* More U.S. children are considered to have elevated blood pressure under new guidelines from the American Academy of Pediatrics.

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Why The Research Is Interesting: The 2017 blood pressure guidelines made two important changes compared to the 2004 fourth report from the National Heart, Lung, and Blood Institute (NHLBI). The first was the exclusion of children who are overweight or obese from the charts used to define normal blood pressures because they tend to have higher blood pressures and worse cardiovascular outcomes later in life. Also, diagnostic thresholds were revised to better align with adult definitions.

Who and When: Fifteen thousand six hundred forty-seven generally healthy, lowrisk children (ages 5 to 18) from the National Health and Nutrition Examination Surveys (NHANES from 1999 to 2014) were used to assess the consequences of the new guidelines classifying blood pressure.

What (Measures): Blood pressure classification based on either the new 2017 American Academy of Pediatrics guidelines or the 2004 NHLBI report.

*How (Study Design):* This was an observational study. Researchers were not intervening for purposes of the study and cannot control for all other factors that could explain the study findings.

*Authors:* Celia J. Rodd, MD, MSc, FRCPC, of the University of Manitoba, Canada, and coauthors.

Study Limitations: Insufficient number of children to assess differences by race, sex and age; a lack of information on medical history, family history and coexisting medical problems; and no long-term follow-up.

(doi:10.1001/jamapediatrics.2018.0223)

http://jamanetwork.com/journals/ jamapediatrics/fullarticle/10.1001/ jamapediatrics.2018.0223

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Gary Webb, MD CHiP Network 215-313-8058 garywebb6@gmail.com



The CHIP Network, the Congenital Heart Professionals Network, is designed to provide a single global list of all CHD-interested professionals.



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