CONGENITAL AORTA TO RIGHT ATRIAL TUNNEL: SUCCESSFUL TRANSCATHETER CLOSURE WITH A DUCTAL OCCLUDER

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Aorticocameral tunnels are extremely rare congenital extracardiac vascular channels, which connect the ascending aorta above the sinutubular junction to any of the chambers of the heart. The ascending aorta is reported to be the most common site of origin but rarely tunnel arising from the descending thoracic aorta has also been reported. More than 90% of the aorticocameral tunnels communicate with the left ventricle, occasionally with the right ventricle, rarely with the atria. The most common of these infrequent conditions is the aorto-left ventricular tunnel (ALVT), followed in frequency by the aorto-right atrial tunnel (ARAT), aorto-right ventricular tunnel (ARVT) and the aorto-left atrial tunnel. All of these conditions produce the physiology of congenital aortic insufficiency, but when the tunnel connects to a right heart chamber, an important left-to-right shunt is also produced.

HISTORY
This is a case of a 20 year old female, born term, with no signs of failure to thrive. At 4 years old, had consult with a private doctor due to recurrent cough, and an incidental murmur was noted. She was managed as a case of Rheumatic Heart Disease with monthly Benzathine Penicillin G IM injections. She was asymptomatic since then with no history of hospitalizations. Until 2 years PTA, patient started to have chest pains and palpitations, but still can do her activities of daily living without difficulty of breathing. With the persistence of chest pain described as squeezing in character, she sought consult at Philippine Heart Center-OPD and several diagnostic work ups were done. She was then advised for coronary angiography. Patient was a nonsmoker and non alcoholic beverage drinker. No history of illicit drug intake. Her current medications were Furosemide 20mg OD, and Enalapril 5 mg OD, with good compliance.

PHYSICAL EXAMINATION
- Ambulatory and not in respiratory distress
- Vital Signs: CR: 70 bpm RR: 18 cpm O2Sat 99% BP: 90/ 60 mmHg Wt 43.7 kg Ht 153 cm BSA 1.36 m²
- Anicteric sclerae, Pink palpebral conjunctivae
- No neck vein engorgement, no carotid bruit
- SCE, no retraction, bronchovesicular breath sounds
- Dynamic precordium, AB 6th ICS LMCL, no thrill/heave, normal rate, regular rhythm, S1 normal, S2 split, grade 3/6 continuous murmur heard best at the right mid parasternal border
- Flat abdomen, soft, nontender, no hepatomegaly, no masses
- No clubbing, no edema, equal peripheral pulses
- Grossly normal extremities

She was admitted at the wards. Complete blood count, protime, aPTT, serum creatinine were within normal limits. There was cardiomegaly with RA and RV prominence on the chest radiograph. Preoperative transthoracic echocardiogram showed anterior type of tunnel from the right coronary sinus to the right atrium.
A hemodynamic study with coronary arteriography was done and revealed angiographically normal coronary arteries. Ascending aortogram showed an anterior type of aorta to right atrium tunnel with a constricted and small opening at the right atrial end.

The closure of an aorta-right atrial tunnel is recommended even in asymptomatic patients as there is only a low rate of procedural complications.

The continued patency of the tunnel leads to risk for biventricular volume overload, bacterial endocarditis, pulmonary vascular disease, aneurysm formation, calcification of the wall, aortic regurgitation and spontaneous rupture.

Treatment options are available according to the type of tunnel, its caliber, tortuosity, calcification, course and relation of the coronary ostia to the aortic orifice of the tunnel. They include:

1. Transcatheter closure
2. Ligation under controlled hypotension or repair with the patient under cardiopulmonary bypass

In this case, a transcatheter treatment is the option of choice due to the small opening of the right atrial end.
PLACEMENT OF THE OCCLUDER DEVICE

- The right femoral vein and artery were cannulated using 6Fr sheaths using standard procedure & the patient was heparinized
- 6F Judkins catheter advanced retrogradely via the ascending aorta and cannulated the ostium of the ARAT from a peripheral access point
- A long Terumo glide wire 0.032 x 260 mm was used to cross the ARAT from the aorta to the RA, RV and to the MPA
- Snare system was inserted antegradely via the right femoral vein up to the MPA. The terumo glide wire was then pulled out by the snare to the right femoral vein
- An arteriovenous guide wire splint was then created
- The CDC6F delivery sheath with introducer was advanced over the wire to the IVC, RA, & to the tunnel
- The dilator & wire were then gently removed
- A 10/12 mm Cocoon PDA device occluder was then deployed at the exit site of the ARAT at the RA side
- Cineangiography post occlusion of ARAT showed the device positioned within the tunnel exit, with minimal shunting of contrast in the center of the device
- Device was then released from the delivery cable

Post intervention, there was no aortic insufficiency noted. The patient tolerated the procedure well and was discharged home after 24 hours of the procedure. A repeat transthoracic echocardiogram was done three months post procedure and showed device in place with total occlusion of the tunnel. There was no aortic insufficiency noted.

CONCLUSION

Aorticocameral tunnels are extremely rare congenital cardiac anomalies. Imaging by TTE, MRI and angiography are of great help in diagnosis. Surgical closure of tunnel along with repair of the associated cardiac defects has been achieved with satisfactory results in the past. However, with the availability of newer innovations and technology, transcatheter closure of tunnels with coils or duct occluders have become a better and more attractive alternative to surgery in selected cases without associated cardiac defects.

REFERENCE