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A transcatheter approach to serial obstructions in an infant with Shone's Complex

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First described in 1963, Shone's Complex is a rare congenital heart lesion comprising at least three serial obstructive lesions of the left side of the heart (1) Supravalvular mitral membrane (2) Parachute mitral valve (3) Muscular or membranous subaortic stenosis and (4) Coarctation of the aorta. Best outcomes have been described with this rare anomaly if the surgical intervention is undertaken before onset of pulmonary hypertension. However, outcomes after transcatheter interventions have rarely been described. We present the case of a 3-month-old baby boy born at term with severe respiratory distress requiring endotracheal intubation at delivery room. Upon evaluation by cardiology noted with supravalvular mitral membrane, subaortic stenosis and severe aortic coarctation consistent with Shone's Complex. A large patent ductus arteriosus (PDA) was also identified for which pharmacologic patency was achieved with alprostadil. Due to patient's critical condition, evidenced by excessive respiratory requirements, a surgical approach was not warranted. A transcatheter approach was considered, upon reasonable clinical stability, both for aortic valvuloplasty and angioplasty as well as device closure of PDA. Interventions were performed with marked improvement in both aortic valve and coarctation gradients but closure had to be deferred to avoid excessive contrast administration. Post-transcatheter course was complicated by right lower extremity occlusion responsive to heparin. He was able to be weaned from mechanical ventilator and remains stable one year after intervention. More than a palliative procedure, this case highlights the possibility of a successful transcatheter approach to the repair of major obstructive defects involving Shone's Complex.

Biography

Guillermo Torres Viera is a Medical Resident in the Combined Internal Medicine-Pediatrics Residency Program of the University of Puerto Rico School of Medicine. He has completed his MD at the University of Puerto Rico in 2014 and entered the first class of this combined specialty program to be created in Puerto Rico for the advancement of knowledge and skill in the effective transition from youth to adulthood. He is currently also pursuing his Masters in Clinical and Translational Research focused towards the identification and prevention of early-onset cardiovascular disease.

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