What to Do for a 75 Year Old Man of Severe Ebstein Anomaly with Decreased RV Function, Atrial Fibrillation

Jong-Min Song, MD, PhD
Cardiology, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea


Recommendations for Surgical Interventions

Class I
Surgeons with training and expertise in CHD should perform tricuspid valve repair or replacement with concomitant closure of an ASD, when present, for patients with Ebstein's anomaly with the following indications:

- Symptoms or deteriorating exercise capacity. (Level of Evidence: B)
- Cyanosis (oxygen saturation less than 90%). (Level of Evidence: B)
- Paradoxical embolism. (Level of Evidence: B)
- Progressive cardiomegaly on chest x-ray. (Level of Evidence: B)
- Progressive RV dilation or reduction of RV systolic function. (Level of Evidence: B)

Recommendations for Catheter Intervention

Class I
Adults with Ebstein's anomaly should have catheterization performed at centers with expertise in catheterization and management of such patients. (Level of Evidence: C)

Class IIa
Catheter ablation can be beneficial for treatment of recurrent supraventricular tachycardia in some patients with Ebstein's anomaly. (Level of Evidence: B)

ESC Guidelines (2010)

Indications for surgery

Class I
Surgical repair should be performed in patients with more than moderate TR and symptoms (NYHA class >II or arrhythmias) or deteriorating exercise capacity measured by CPET (Level of Evidence: C)
If there is also an indication for tricuspid valve surgery, then ASD/PFO closure should be performed surgically at the time of valve repair (Level of Evidence: C)

Class IIa
Surgical repair should be considered regardless of symptoms in patients with progressive right heart dilation or reduction of RV systolic function and/or progressive cardiomegaly on chest X-ray (Level of Evidence: C)

Indications for catheter intervention

Class I
Patients with relevant arrhythmias should undergo electrophysiologic testing, followed by ablation therapy, if feasible, or surgical treatment of the arrhythmias in the case of planned heart surgery (Level of Evidence: C)

Class IIa
In the case of documented systemic embolism probably caused by paradoxical embolism, isolated device closure of ASD/PFO should be considered (Level of Evidence: C)

Class IIb
If cyanosis (oxygen saturation at rest <90%) is the leading problem, isolated device closure of ASD/PFO may be considered but requires careful evaluation before intervention (Level of Evidence: C)