# WCC 2016-107: Unusual Presentation of Ebstein's Anomaly – Stenotic Variant Presenting with Paradoxical Embolism in Adult Female

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**Introduction:**

Ebstein's anomaly of tricuspid valve, an embryological defect of delamination of inlet of right ventricle occurs in approximately 1 in 20,000 live births, accounts for 0.3% to 0.7% of all cases of congenital heart disease, and represents about 40% of congenital malformations of the tricuspid valve. It is characterized by variable apical displacement of septal, posterior leaflets of the tricuspid valve by more than 8mm/m2.It is unique for its varied types of presentations at different stages of life, ranging from fetus to elderly. Stenotic variant of ebstein's anomaly is very rare subgroup, here we are reporting a rare case presenting with paradoxical embolism.

**Case:**

18 year old female, previously known case of congenital heart disease, presented with painful swelling of both lower limbs since one month, dyspnea on exertion NYHA class 2 and moderate cyanosis. she was admitted and evaluated. Investigations revealed thrombosis of left popliteal artery, with stenotic ebsteins anomaly of tricuspid valve, large OS ASD ,right to left shunt, large atrialised RV, small functional right ventricle with good sized confluent pulmonary arteries (type D carpentier).she was treated conservatively with anticoagulants for the peripheral artery thrombosis. Catheterization was done, which revealed the small sized functional RV with good sized confluent pulmonary arteries, right to left shunt of 2.2:1 across the ASD and anomalous origin of RCA from left coronary sinus. Patient was sent for surgery with bidirectional glenn shunt, tricuspid valve repair and closure of ASD after 6 weeks of anticoagulation. Her postoperative course was uneventful and was discharged after 2 weeks of hospital stay.

**Discussion:**

Ebstein's anomaly of tricuspid valve – stenotic variant is a rare entity with only few isolated case reports. Paradoxical embolism is a known complication in ebstein's anomaly and is an indication for surgery in these patients. The rarity of the variant makes it worthwhile to know for cardiologist treating congenital heart disease.

**Conclusion**:

Ebstein's anomaly is a complex congenital anomaly with a broad anatomic and clinical spectrum. Management is complex and must be individualized. Precise knowledge about the different anatomic and hemodynamic variables, associated malformations, and management options is essential.

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