

## **Management of Intra-Capsular Fracture Neck of Femur in Case of Ebstein's Anomaly**

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### **Abstract**

Ebstein's anomaly is a rare congenital heart disease, which involves downward displacement of tricuspid valve and atrialisation of right ventricle associated with either atrial septal defect or patent foramen ovale through which shunting of blood occurs. In such patient, clinical presentation varies from congestive cardiac failure in childhood to incidental diagnosis in adulthood. If such patient presents with fracture neck of femur, it will be a challenging job for orthopaedic surgeon as well as anaesthesiologist. The main goals of treatment are to stabilize the hip, decrease pain and restore the level of pre-fracture function with minimal operative intervention.

**Keywords:** Ebstein's anomaly; intra-capsular femur neck fractures; hemi-replacement arthroplasty; avascular necrosis; epidural anaesthesia.

### **Introduction**

Hip fractures are common in a frail elderly patients which can be difficult to manage. The mortality rate is in the 20% to 24% range at 1 year; many patients will lose their independence after hip fracture. Ebstein's anomaly is an abnormality of tricuspid valve in which valve leaflets are displaced downwards into right ventricle, resulting in large right atrium, small right ventricle and tricuspid regurgitation or stenosis [1]. It occurs in 1 percent of congenital heart defects [2]. The incidence in general population is 1:110,000 and there is no sex difference. This anomaly was described by Wilhelm Ebstein in 1866 in a report titled, "Concerning a very rare case of insufficiency of the tricuspid valve caused by a congenital malformation"[3]. It is associated with either atrial septal defect (septum secundum) or patent foramen ovale through which shunting of blood occurs. Haemodynamic derangement depends upon status

of tricuspid valve displacement and right to left shunt. Patients with fracture neck of femur with associated Ebstein's anomaly have to be addressed with a proper approach for definitive fracture management keeping in mind the deleterious effects of uncorrected congenital heart defect on health of the patient.

### **Case Report**

A 52 year old male with history of fall from bike due to skid 3 days back at presented at Emergency Department with complaints of pain in left hip, inability to bear weight on left lower limb with painful and restricted movements of the left hip joint. Moreover, patient gave history of being a cardiac patient suffering from congenital heart disease - Ebstein's anomaly. He had exertional dyspnoea (Grade 2 NYHA) since 15 years due to which he limited his activities. He was on treatment with Digoxin and Torsemide with Spironolactone.

However patient was not compliant with these medications.



Fig. 1: Pre-Op X-ray of pelvis with both hips (AP view).

On examination, patient was conscious and well-oriented with *Pulse Rate* - 112 bpm in right radial pulse, no radio-radial delay, radio-femoral delay, *BP*-130/80 mmHg in both arms in supine position. On systemic examination of cardiovascular system- S1S2 were heard with ESM (ejection systolic murmur) in pulmonary area (Grade 2), and in respiratory system- bilateral air entry was present with no added sounds.

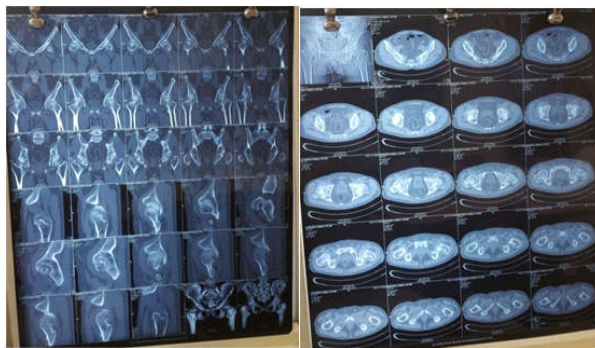


Fig. 2: Pre-Op CT Scan of pelvis with both hips

On local examination, swelling and tenderness were present over left hip with restricted range of movements of left hip with left lower limb externally rotated. Pelvic compression test was positive. Neurovascular examination was unremarkable and Ankle-Brachial index was 1.0 on the injured leg. On further radiological evaluation, X-rays revealed trans-cervical (trans-cervical) type of neck of femur fracture, with no other bony injuries. When subjected to CT scanning, it was found that it was in fact a comminuted fracture of neck of femur (trans-cervical). Patient's basal oxygen saturation was 90% on an average (85-92%), which



Fig. 3: Post-Op X-ray of pelvis with both hips with implant in situ (AP view).

was improved upto 94% with 4 to 8 L of oxygen per min. Cardiology opinion and pre-operative fitness was taken to optimize patient's cardiac condition. Pre-operative 2D ECHO was suggestive of -situs solitus with D-Loop Ventricle, acyanotic congenital heart disease, Ebstein' anomaly of the tricuspid valve, apical displacement of tricuspid septal leaflet, atrialised right ventricle and dilated right atrium, intact IAS/IVS, aortic valve thickened with trivial AR, grade 1 TR. With grave-risk consent, uncemented bipolar hemiarthroplasty of neck of femur fracture was performed through antero-lateral approach under epidural anaesthesia with adequate analgesia. Throughout the surgery patient his vital signs were unchanged to any significant degree with oxygen saturation being 85% with 100% Oxygen @ 10L/min by face-mask. post operative period was un-eventful except for the patient's oxygen saturation that was 85 to 90% with O2 mask which gradually improved over 3 days and patient maintained the 90% saturation without O2 mask. Gradual passive ROM exercises were started within 2 days with partial to full weight bearing mobilization with a walker over a period of 2 weeks. With active physiotherapy sessions during follow up, the patient progressed to full weight bearing without assisted device by 4 weeks post-operatively with satisfactory range of motion at left hip. Patient is satisfied with his function and stability, and feels adequate to perform job related activities and activities of daily living.



Fig. 4: follow up at 4 weeks- patient able to stand, walk, sit crossed legs, squat, do SLR.

## Discussion

In patient with Ebstein's anomaly, disease severity depends upon the degree of valvular abnormality, direction of intra-cardiac shunting, pulmonary hypertension, ventricular and supra-ventricular tachycardia and association with Wolf-Parkinson-White syndrome. These patients are at risk of paradoxical embolism, brain abscess, CCF and sudden death [4]. Similarly, in our case septum secundum ASD was present in the previous echocardiography which had got resolved over the years probably, hence not detected in preoperative ECHO. Moreover, the intraoperative findings of low oxygen saturation did not correlate with the pre-operative 2D-Echo findings of absent ASD and PAH. If such patient comes for emergency surgery, anaesthesia technique is to be optimized to avoid worsening of the pre-existing condition, that is to avoid decrease in systemic vascular resistance or increase in pulmonary resistance. In this patient it was bidirectional shunt, mainly left to right shunt. Because of that he was asymptomatic. The main aim was to maintain left to right shunt and avoid reversal of shunt. During general anaesthesia, increased intrathoracic pressure due to positive pressure ventilation aggravates right to left shunt and also increased sympathetic response during intubation and extubation which can precipitate adverse cardiac events, and so it is not preferred. To avoid complication of general anaesthesia and to provide benefits of regional anaesthesia to clinically stable patient for bipolar hemi-arthroplasty we preferred epidural anaesthesia to minimize hemodynamic changes which cause an increase in right-to-left shunt and hypoxemia. Fractures involving the femoral neck can disrupt the vascular

supply to the femoral head and result in avascular necrosis (AVN) or nonunion [5]. A meta-analysis of the outcome of displaced femoral neck fractures found the rates of osteonecrosis and nonunion to be as high as 20% to 30% [5]. Surgical stabilization should be performed as soon as possible—ideally, within 48 hours to achieve primary osteosynthesis. In delayed presentation following injury in hip fractures hemi or total hip replacement is considered to be better option, to avoid risk of further avascular necrosis and need for revision surgeries in high risk elderly patients. Given the complexity of his congenital cardiac anomaly with a relatively normal acetabulum, the patient was treated with bipolar hemi-arthroplasty instead of extensive surgical intervention like total hip replacement.

## Conclusion

The successful management of a patients with Ebstein's anomaly requires a deep understanding of the pathophysiology of this cardiovascular disease and its effects on bone healing. Such patient should be considered as high risk cases and must be managed in tertiary centres by a multidisciplinary team. If patient is clinically stable and optimised, minimal needed intervention i.e. bipolar hemi-arthroplasty under epidural anaesthesia provides better option for fracture neck of femur cases.

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