From ultrasound to intervention: Indomethacin in antenatal management of Ebstein anomaly

Chai Hong Lim[1], Wang Ching Chua[1], Vairavan Ramesh V[1], Putri Yubbu[2]

[1]Department of Obstetrics and Gynaecology, Hospital Sultan Idris Shah, Serdang, Selangor, Malaysia, [2]Department of Paediatric Cardiology, Hospital Sultan Abdul Aziz Shah, UPM, Selangor, Malaysia

ABSTRACT

Introduction: Ebstein anomaly (EA) is a rare congenital heart defect characterized by the apical displacement of the tricuspid valve. The severity of EA is classified using the GOSE score. Factors associated with poorer outcomes include fetal hydrops and circular circulation, the latter of which involves pulmonary regurgitation and excessive flow across the ductus arteriosus. Indomethacin, a non-steroidal anti-inflammatory drug, is known to constrict the ductus arteriosus but may also adversely affect fetal urine production. **Case Description:** A 27-year-old woman in her second pregnancy was seen at 27 weeks gestation with an anomaly scan revealing a fetus with EA. The family history was unremarkable, and the patient was taking T. Sertraline 50 mg nightly for depression. A fetal echocardiogram performed at 29 weeks showed moderate EA with severe tricuspid regurgitation. By 33 weeks, the condition had progressed to include pulmonary regurgitation and hydrops, evidenced by pericardial effusion, ascites and polyhydramnios. Reversal of flow was noted at the ductal arch, with a large, tortuous patent ductus arteriosus. A multidisciplinary discussion involving fetal medicine and perinatal cardiologists led to the decision to start Indomethacin at 100 mg twice daily at 33 weeks, which was then tapered to 25 mg four times daily. A follow-up scan at 34 weeks showed no obvious reversal of ductal flow, resolution of ascites and a normal amniotic fluid index until delivery. The patient delivered via cesarean section at 37 weeks. **Discussion:** Prenatal Indomethacin therapy successfully constricted the ductus arteriosus, resolving the circular shunt and hydrops, thereby averting severe morbidity and mortality. Close dose adjustments were necessary to avoid anhydramnios and fetal renal injury.