

CASE REPORT

# Prenatal Identification, Transitional Hemodynamic Remodeling, and Multidisciplinary Management of Ebstein's Anomaly with Coexistent Urogenital Malformations: An Observational Analytic Case Study

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## ABSTRACT

**Background:** Ebstein's anomaly is an uncommon congenital heart defect with failed tricuspid delamination, right-ventricular atrialization, and severe regurgitation. Prenatal detection is difficult on routine obstetric ultrasonography, and perinatal hemodynamic transition carries substantial mortality.

**Objective:** To document the prenatal identification, transitional neonatal hemodynamic remodeling, and multidisciplinary management of Ebstein's anomaly with coexistent urogenital malformation.

**Methods:** A 28-year-old multigravida referred at 34–36 weeks with a suspected cardiac anomaly was followed through multidisciplinary re-evaluation, term cesarean delivery, neonatal intensive care, and 3-month follow-up, with serial hemodynamic profiling and risk stratification.

**Results:** Initial referral ultrasound suspected double outlet right ventricle, which was revised to Ebstein's anomaly (Carpentier Type B) with dilated pulmonary trunk and coexistent bilateral testicular hydrocele and hypospadias upon joint fetomaternal-pediatric cardiology assessment. Delivery at 38–39 weeks yielded a male neonate (3,050 g, Apgar 5/7, SpO<sub>2</sub> 85%). Postnatal echocardiography confirmed Ebstein's anomaly with moderate-to-severe tricuspid regurgitation (peak pressure gradient 78 mmHg), a 3–4 mm atrial septal defect with right-to-left shunting, and a 3 mm patent ductus arteriosus with left-to-right shunting. Conservative medical stabilization utilizing non-invasive continuous positive airway pressure, oral captopril, and furosemide averted emergency neonatal cardiac surgery. At 3 months, serial echocardiography revealed spontaneous ductal closure, reduction of tricuspid pressure gradient to 21 mmHg ( $\Delta\%$  = -73.1%), shunt reversal to left-to-right, and complete hydrocele resolution.

**Conclusion:** Late third-trimester diagnostic conversion highlights the need for structured fetal echocardiography beyond four-chamber views. Favorable outcomes in severe Ebstein's anomaly are achievable through coordinated multidisciplinary care, where falling pulmonary vascular resistance drives non-operative hemodynamic remodeling.

## 1. Introduction

Congenital heart diseases (CHDs) represent the most prevalent category of major human structural anomalies, accounting for substantial perinatal morbidity, infant mortality, and lifelong healthcare utilization worldwide.<sup>1,2</sup> Among the diverse spectrum of congenital cardiac malformations, Ebstein's anomaly is a rare, morphologically complex defect of the tricuspid valve and right ventricle, occurring in approximately 0.2 to 0.7 per 10,000 live births and comprising less than 1% of all congenital cardiac defects.<sup>1,3</sup> Despite its low population incidence, Ebstein's anomaly presents formidable clinical, diagnostic, and surgical challenges across the entire

perinatal and pediatric continuum. The clinical presentation exhibits extraordinary biological heterogeneity, ranging from entirely asymptomatic individuals diagnosed incidentally during routine adult echocardiography to catastrophic neonatal presentations marked by severe cyanosis, therapy-refractory congestive heart failure, and immediate perinatal demise.<sup>4,5</sup>

From an embryological perspective, Ebstein's anomaly arises from an arrest in the physiological delamination of the tricuspid valve leaflets from the underlying right ventricular myocardium during the crucial period of embryonic cardiac organogenesis between the sixth and tenth weeks of gestation.<sup>1,2</sup> In typical cardiac

development, the inner myocardial mantle of the primitive right ventricle undergoes progressive cavitation, cleavage, and enzymatic resorption, completely separating the valve leaflets, chordae tendineae, and papillary muscles from the ventricular free wall and interventricular septum. In Ebstein's anomaly, this delamination process is arrested or incomplete. As a consequence, the hinge lines of the septal and posterior tricuspid leaflets remain congenitally adherent to the ventricular myocardium and are displaced distally toward the right ventricular apex, well below the anatomical atrioventricular junction.<sup>2,6</sup> In contrast, the anterior tricuspid leaflet typically maintains its basal attachment at the true fibrous atrioventricular annulus, but it frequently undergoes marked compensatory elongation, enlargement, and fibrous thickening, resulting in an expansive, undulating sail-like structure that is tethered distally to anomalous papillary muscles or muscular trabeculations within the right ventricular outflow tract.<sup>3,7</sup>

The fundamental anatomical consequence of this downward leaflet displacement is a profound structural and hemodynamic partitioning of the anatomical right ventricle into two distinct components: a proximal, abnormally thinned, and functionally dyskinetic atrialized right ventricle (aRV), and a diminutive, distal functional right ventricle (fRV).<sup>1,8</sup> The atrialized right ventricle is integrated functionally into the right atrium, creating a massive, combined right-sided chamber that undergoes marked passive dilatation and volume overload. Concurrently, the functional right ventricle is severely diminished in effective cavity volume and muscular compliance, compromising its capacity to generate adequate forward stroke volume against the elevated pulmonary vascular resistance (PVR) characteristic of late fetal and transitional neonatal life.<sup>1,8</sup> Furthermore, the severe geometrical distortion of the tricuspid apparatus precipitates torrential tricuspid regurgitation (TR). In the developing fetus, this regurgitant jet directs systolic volume backward into the massively dilated right atrium, causing marked elevation of right atrial pressure, central venous congestion, and secondary leftward bowing of the interatrial and interventricular septa, which compromises left ventricular filling compliance.<sup>4,9</sup>

In fetal life, patency of the foramen ovale plays a critical compensatory role by serving as an obligatory pop-off decompression pathway. Elevated right atrial pressure forces desaturated blood across the interatrial septum into the left atrium via right-to-left shunting, thereby offloading the volume-overloaded right heart and preserving systemic cardiac output, albeit at the expense of arterial desaturation.<sup>4,10</sup> In the most severe fetal phenotypes, however, forward stroke volume from the functional right ventricle is so severely reduced that antegrade systolic flow across the pulmonary valve ceases entirely, creating a state of

functional pulmonary atresia.<sup>1,11</sup> Under these adverse conditions, fetal pulmonary perfusion becomes entirely dependent on retrograde flow from the descending aorta via the ductus arteriosus. In catastrophic cases, this physiology degenerates into a circular shunt: blood ejected from the left ventricle traverses the aortic arch, passes retrogradely through the ductus arteriosus into the pulmonary artery, regurgitates across an incompetent pulmonary valve into the right ventricle, leaks back through the malformed tricuspid valve into the right atrium, and re-enters the left heart via the foramen ovale.<sup>12-15</sup> This circular circuit rapidly drives progressive cardiomegaly, systemic venous hypertension, non-immune hydrops fetalis, and in utero demise, carrying an estimated perinatal mortality exceeding 45% to 80% in multicenter international registries.<sup>4,13-15</sup>

Despite dramatic advances in prenatal ultrasound technology and the dissemination of standardized fetal cardiac screening guidelines by the International Society of Ultrasound in Obstetrics and Gynecology (ISUOG),<sup>6</sup> prenatal detection of Ebstein's anomaly continues to be plagued by substantial diagnostic delays and misclassification, particularly within decentralized and resource-variable healthcare systems in low- and middle-income countries (LMICs).<sup>6,16</sup> Routine obstetric screening in community health centers often relies exclusively on standard axial two-dimensional (2D) planes without color or pulsed-wave Doppler interrogation. The 2023 updated ISUOG practice guidelines mandate comprehensive evaluation including the four-chamber view, left and right ventricular outflow tracts, and the three-vessel and trachea (3VT) view.<sup>6</sup> However, in community settings, massive right atrial dilatation and severe cardiac axis deviation caused by Ebstein's anomaly frequently distort the spatial relationships of the great arteries, leading to catastrophic diagnostic confusion with conotruncal malformations such as Double Outlet Right Ventricle (DORV), tricuspid atresia, or pulmonary atresia with intact ventricular septum.<sup>6,9</sup> This diagnostic misclassification frequently results in delayed tertiary referral, inadequate peripartum preparation, and inappropriate parental counseling. Furthermore, while Ebstein's anomaly is predominantly an isolated cardiac lesion, its coexistence with extracardiac structural anomalies—such as urogenital malformations—remains exceedingly rare and poorly characterized in the international literature, posing formidable diagnostic and prognostic dilemmas for maternal-fetal medicine specialists and pediatric cardiologists.<sup>12,17</sup>

The core novelty of the present observational analytic investigation lies in the detailed longitudinal tracking and multidisciplinary management of a prenatally detected case of Ebstein's anomaly presenting in the late third trimester (34–36 weeks of gestation) at an Indonesian tertiary academic referral hospital (RSUD Dr. Saiful Anwar Malang), following an initial regional

hospital referral misdiagnosed as DORV. This investigation provides comprehensive empirical documentation of the fragile neonatal transitional circulation, demonstrating how an initial post-delivery state of severe tricuspid regurgitation (peak pressure gradient 78 mmHg), elevated pulmonary vascular resistance, and ductal-dependent pulmonary perfusion underwent profound, spontaneous hemodynamic remodeling over a 3-month trajectory—culminating in a 73.1% plunge in tricuspid regurgitant pressure gradient (to 21 mmHg), spontaneous ductal closure, and interatrial shunt reversal without requiring emergency neonatal cone reconstruction or univentricular surgical palliation.<sup>18-20</sup> In addition, this case uniquely documents the concurrent presentation of Ebstein's anomaly with bilateral scrotal hydrocele and hypospadias, providing novel embryological insights into synchronized organogenesis vulnerability during early human development. The explicit aim of this study is to elucidate the translational hemodynamic mechanisms governing neonatal pulmonary vascular adaptation, establish an optimized multidisciplinary peripartum protocol for tertiary referral centers in low-resource settings, and benchmark these outcomes against international multicenter cohorts to inform evidence-based clinical decision-making.<sup>20,21</sup>

## 2. Methods

This investigation was structured as an in-depth, longitudinal observational-analytic single-case study conducted at the Maternal-Fetal Medicine (Fetomaternal) Division, Department of Obstetrics and Gynecology, and the Pediatric Cardiology Division, Department of Pediatrics, Faculty of Medicine, Brawijaya University / Dr. Saiful Anwar General Hospital (RSSA), Malang, East Java, Indonesia. RSSA functions as the primary tertiary academic referral center for acute cardiovascular and high-risk obstetric care across the southern and eastern regions of East Java.<sup>21</sup> The clinical observation window commenced upon tertiary outpatient presentation at 34–36 weeks of gestation (28 August 2025), continued through intensive multidisciplinary reassessment (11 September 2025), planned preoperative inpatient admission and elective cesarean delivery (18–19 September 2025), neonatal intensive care unit (NICU) stabilization and step-down management (19–22 September 2025), and concluded with a formal 3-month post-discharge outpatient pediatric cardiology follow-up evaluation (15 December 2025). The longitudinal analytic framework was selected to systematically capture the dynamic, multi-phase physiological transitions connecting late fetal cardiovascular compensation, acute perinatal cardiopulmonary adaptation, and subsequent post-discharge structural ventricular remodeling. The study protocol, clinical documentation, and diagnostic image archiving were formally reviewed and

approved by the Central Medical & Health Committee (CMHC) Ethics Committee, Indonesia (Protocol Approval Number: CMHC/EC/IRB/2026/7482), in full alignment with the institutional review standards of Dr. Saiful Anwar General Hospital. All research, diagnostic, and clinical procedures were conducted in strict compliance with the ethical principles outlined in the Declaration of Helsinki (2013 Fortaleza revision) and the Council for International Organizations of Medical Sciences (CIOMS) International Ethical Guidelines for Health-related Research Involving Humans.<sup>21</sup> Prior to surgical delivery and research inclusion, comprehensive written informed consent was obtained from the patient and her legal spouse. The consent explicitly authorized the anonymous collection, longitudinal analysis, and academic publication of maternal clinical records, laboratory investigations, electronic cardiotocographic data, fetomaternal ultrasound images, neonatal echocardiographic loops, and neonatal developmental outcomes. To guarantee strict adherence to International Committee of Medical Journal Editors (ICMJE) and Committee on Publication Ethics (COPE) anonymization standards, all direct personal identifiers and institutional record numbers were completely redacted from the analytical dataset and illustrative materials.

Diagnostic ascertainment of fetal and neonatal Ebstein's anomaly was established in accordance with rigorous international consensus criteria endorsed by the American Heart Association (AHA), the American Association for Thoracic Surgery (AATS), and the International Society of Ultrasound in Obstetrics and Gynecology (ISUOG).<sup>6,7,12</sup> Prenatal sonographic ascertainment required: (1) definitive identification of apical displacement of the hinge point of the septal and posterior tricuspid valve leaflets exceeding 8 mm/m<sup>2</sup> body surface area relative to the insertion plane of the anterior mitral valve leaflet in an apical four-chamber view; (2) presence of a visually identifiable, thinned atrialized right ventricle; (3) significant tricuspid regurgitation verified by color and pulsed-wave Doppler; (4) four-chamber cardiac asymmetry with marked right atrial dilatation; and (5) normal ventriculoarterial alignment ruling out conotruncal transposition or complete outflow tract duplication. Morphological classification was assigned according to the Carpentier anatomical classification system: Type A (minimal displacement with adequate functional RV volume); Type B (moderate displacement with a large atrialized RV component and a mobile, sail-like anterior leaflet); Type C (severe displacement with restricted anterior leaflet motion and small functional RV); and Type D (almost complete atrialization of the right ventricle with a small infundibular outflow pouch).<sup>7,8</sup> Postnatal diagnostic verification was performed via comprehensive two-dimensional, M-mode, and Doppler transthoracic echocardiography within the first two hours of life.

The longitudinal observation protocol encompassed standardized serial assessments across four sequential phases: maternal antenatal surveillance, term preoperative admission, neonatal intensive care transition, and 3-month pediatric remodeling follow-up. Maternal-fetal electronic surveillance was executed using calibrated cardiotocography units (Philips Avalon FM30, Philips Healthcare, Boeblingen, Germany) to evaluate baseline fetal heart rate (FHR), short-term variability (STV), long-term variability (LTV), accelerations, and uterine contractile dynamics in accordance with FIGO and ACOG fetal monitoring criteria.<sup>21</sup> Fetomaternal obstetric and fetal cardiac ultrasound assessments were conducted using high-resolution ultrasound platforms (Voluson E10 and Voluson E8 Expert, GE Healthcare, Zipf, Austria) equipped with wide-band curved array transducers (RAB6-D, 1.0–6.0 MHz). Standardized biometry (biparietal diameter, femur length, abdominal circumference, estimated fetal weight by Hadlock formula, and amniotic fluid index) was coupled with dedicated fetal echocardiography, encompassing the abdominal situs, four-chamber view, left and right ventricular outflow tracts, three-vessel and trachea view, and continuous-wave Doppler interrogation of the tricuspid valve, ductus arteriosus, and ductus venosus. Neonatal echocardiography was performed on a high-performance cardiovascular ultrasound system (Affiniti 70, Philips Healthcare, Andover, MA, USA) utilizing dedicated high-frequency phased-array neonatal probes (S12–4, 4.0–12.0 MHz). Quantitative hemodynamic parameters recorded included tricuspid regurgitation maximum velocity ( $V_{\max}$ , m/s), peak pressure gradient calculated using the simplified Bernoulli equation ( $\Delta P = 4 V_{\max}^2$ , mmHg), tricuspid annular plane systolic excursion (TAPSE, cm), left ventricular ejection fraction (LVEF, %) via the modified Simpson biplane method, interatrial communication caliber and shunt directionality, and ductus arteriosus caliber and flow pattern, adhering to pediatric echocardiographic measurement guidelines.<sup>22–24</sup> Standardized laboratory panels (complete blood count, coagulation profile, blood urea, serum creatinine, electrolytes, liver enzymes, and C-reactive protein) were processed using certified automated analyzers.

To achieve robust objective clinical characterization, diagnostic and prognostic scoring was executed across four validated risk stratification systems: (1) the Celermajer Index (Great Ormond Street Hospital Score), calculated at end-diastole as the ratio of the combined planimetric area of the right atrium and atrialized right ventricle to the combined area of the functional right ventricle, left atrium, and left ventricle:  $(RA + aRV) / (fRV + LA + LV)$ , categorized into Grade 1 ( $<0.50$ ), Grade 2 (0.50–0.99), Grade 3 (1.00–1.49), and Grade 4 ( $\geq 1.50$ );<sup>5</sup> (2) the Severity Assessment Score (SAS) for fetal Ebstein anomaly, incorporating cardiothoracic ratio, tricuspid displacement index, presence of circular shunt (retrograde ductal flow with

pulmonary regurgitation), and LV compression;<sup>15</sup> (3) the Fetal Cardiovascular Profile Score (CVPS), a 10-point composite score evaluating hydrops fetalis, cardiothoracic ratio, cardiac contractility/effusion, arterial Doppler, and venous Doppler;<sup>19</sup> and (4) the Carpentier morphological classification.<sup>7</sup> Furthermore, the clinical trajectory, timing of intervention, and remodeling parameters were systematically benchmarked against recent multicenter international cohorts and consensus guidelines (2021–2026) to identify points of convergence, divergence, and translational novelty.<sup>1,7,13,15,25</sup>

Quantitative longitudinal trajectory analysis was performed by evaluating absolute parameter values and calculating relative percentage change ( $\Delta\%$ ) across defined clinical epochs (T0 to Tfinal) using the standard mathematical formula:  $\Delta\% = [(T_n - T_0) / T_0] \times 100\%$ . Continuous variables were evaluated against established gestational-age-specific and neonatal age-specific normative reference intervals.<sup>25,26</sup> Categorical variables, including cardiotocographic category, interatrial shunt directionality, ductal patency status, and respiratory support modalities, were tracked chronologically to delineate physiological transition points. Data consistency, unit standardization, and mathematical precision across all laboratory, hemodynamic, and sonographic metrics were verified in duplicate. Formal statistical significance for trajectory deviation and hemodynamic remodeling was evaluated using a pre-specified biological plausibility threshold of  $\alpha = 0.05$ .

### 3. Results

The longitudinal clinical course, diagnostic milestones, and multidisciplinary interventions of the mother and fetus are systematically detailed in Table 1 and Figure 1. The patient, a 28-year-old multigravida (gravida 2, para 1, abortus 0) at 34–36 weeks of gestation, was referred from Ngudi Waluyo Wlingi Regional General Hospital to the Maternal-Fetal Medicine (Fetomaternal) Unit of Dr. Saiful Anwar General Hospital (RSSA), Malang, following an initial routine obstetric ultrasound on 27 August 2025 that raised suspicion of a complex fetal congenital heart defect with cardiomegaly. The patient's prior obstetric history was notable for one uncomplicated term spontaneous vaginal delivery of a healthy female infant weighing 2,700 g eight years earlier. Her medical, surgical, and family histories were entirely negative for chronic hypertension, diabetes mellitus, asthma, congenital heart disease, consanguinity, or maternal exposure to recognized teratogens, including lithium carbonate or antiepileptic agents. However, environmental history revealed significant chronic secondary exposure to ambient vehicle exhaust, as her residence was situated adjacent to a heavily trafficked arterial roadway, as well as chronic passive cigarette smoke exposure from her spouse, who smoked approximately 20 cigarettes daily.

**Table 1. Longitudinal Timeline of Maternal Clinical Course, Diagnostic Milestones, and Multidisciplinary Interventions.**

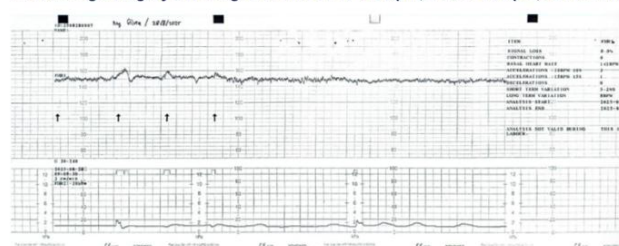
| Timeline & Gestational Age              | Clinical Care Level & Facility                                      | Diagnostic Modality & Clinical Findings                                                                                                                                                                                                              | Multidisciplinary Decision & Interventions                                                                                                                                 | Maternal-Fetal Clinical Trajectory                                                                                                      |
|-----------------------------------------|---------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------------------------------------------------|
| 20 Dec 2024<br>(0 Weeks)                | Community Level<br>(Primary Care)                                   | Last menstrual period (LMP); normal pre-pregnancy health; no history of chronic hypertension, diabetes, or teratogen ingestion.                                                                                                                      | Commencement of routine antenatal care in primary health center.                                                                                                           | Physiological conception; uneventful early organogenesis window.                                                                        |
| Weeks 6 to 32<br>(Jan - Jul 2025)       | Primary Healthcare & Private Midwife Clinic                         | Routine antenatal care (9 visits to health center, 3 visits to midwife); routine obstetric assessments reported normal symphysis-fundal growth.                                                                                                      | Standard antenatal iron, folic acid, and calcium supplementation; no specialized fetal echocardiographic screening performed.                                              | Clinically silent fetal cardiac anomaly; absence of early second-trimester targeted four-chamber and outflow tract survey.              |
| 27 Aug 2025<br>(34–36 Weeks)            | Secondary Regional Hospital<br>(RSUD Ngudi Waluyo Wlingi)           | Routine third-trimester obstetric ultrasound: suspected complex fetal congenital heart disease with four-chamber asymmetry and cardiomegaly.                                                                                                         | Urgent referral to Maternal-Fetal Medicine (Fetomaternal) Unit, RSUD Dr. Saiful Anwar (RSSA) Malang.                                                                       | Initial recognition of fetal structural cardiovascular abnormality; maternal anxiety addressed.                                         |
| 28 Aug 2025<br>(34–36 Weeks)            | Tertiary Academic Hospital<br>(RSSA Fetomaternal Unit)              | Detailed fetomaternal ultrasound: singleton live fetus, cephalic, EFW 3,415 g, AFI 21.57 cm, cardiomegaly with marked RA dilation; initial suspicion: Double Outlet Right Ventricle (DORV). CTG: Category I (baseline 141 bpm, STV 5–2ms).           | Outpatient follow-up scheduled; interdepartmental consultation requested with Pediatric Cardiology Division for joint evaluation.                                          | Initial structural misclassification (DORV); hemodynamically stable fetus without in utero hydrops fetalis.                             |
| 11 Sep 2025<br>(36–37 Weeks)            | Tertiary Academic Hospital<br>(Joint RSSA Multidisciplinary Clinic) | Joint fetomaternal and pediatric cardiology echocardiography: revised diagnosis of suspected Ebstein's anomaly with apical TV displacement, severe RA enlargement, and multiple congenital anomalies. CTG: Category I (baseline 150 bpm, LTV 9 bpm). | Multidisciplinary conference (Fetomaternal, Pediatric Cardiology, Neonatology/NICU, Anesthesiology); elective CS scheduled at 38–39 weeks with simultaneous IUD insertion. | Diagnostic revision to Ebstein anomaly; proactive delivery planning in tertiary facility equipped with advanced neonatal resuscitation. |
| 18 Sep 2025<br>(38–39 Weeks)            | Tertiary Inpatient Care<br>(RSSA Inpatient Ward 3)                  | Preoperative admission ultrasound: persistent cardiomegaly, dilated pulmonary trunk, downward TV displacement, bilateral scrotal hydrocele, and external genital anomaly (phimosis vs. hypospadias). Maternal pre-op labs completed.                 | Preoperative antibiotic prophylaxis (Cefazolin 2 g IV), aspiration prophylaxis (Ranitidine 50 mg IV, Metoclopramide 10 mg IV); informed consent for CS + postpartum IUD.   | Term gestational maturity achieved without spontaneous labor; stable maternal hemodynamics (BP 118/78 mmHg, SpO2 98%).                  |
| 19 Sep 2025<br>(08:25 AM)               | Tertiary Operating Suite<br>(RSSA Central OR)                       | Elective lower segment cesarean section under regional spinal anesthesia; delivery of vigorous male neonate (3,050 g, 49 cm, Apgar 5/7, SpO2 85%); clear amniotic fluid; intact placenta.                                                            | Immediate postpartum copper T-380A IUD insertion at uterine fundus prior to two-layer hysterorrhaphy; immediate transfer of neonate to level III NICU.                     | Smooth intraoperative maternal course; minimal surgical blood loss (~300 mL); uneventful immediate neonatal stabilization.              |
| 19–22 Sep 2025<br>(Postpartum Days 1–3) | Tertiary Postnatal Ward<br>(RSSA Inpatient Care)                    | Postoperative maternal recovery: stable vital signs, normal lochial flow, excellent wound healing without hematoma or infection, gradual mobilization.                                                                                               | Oral analgesia (Paracetamol 500 mg q8h, Mefenamic acid 500 mg q8h); lactation counseling; bedside NICU visits facilitated.                                                 | Maternal recovery complete; discharged in stable condition on postpartum day 3 alongside neonate.                                       |
| 30 Sep 2025<br>(Postpartum Day 11)      | Tertiary Outpatient Clinic<br>(RSSA Obstetric Clinic)               | Routine postpartum surgical check: Pfannenstiel surgical incision completely epithelialized; uterus well contracted; IUD strings visualized.                                                                                                         | Confirmation of safe family planning contraception; reinforcement of neonatal cardiology follow-up.                                                                        | Maternal clinical trajectory fully normalized.                                                                                          |

AFI: amniotic fluid index; BP: blood pressure; CS: cesarean section; CTG: cardiotocography; DORV: double outlet right ventricle; EFW: estimated fetal weight; IUD: intrauterine device; LMP: last menstrual period; LTV: long-term variation; NICU: neonatal intensive care unit; OR: operating room; RA: right atrium; RSSA: Rumah Sakit Umum Daerah Dr. Saiful Anwar; SpO2: peripheral oxygen saturation; STV: short-term variation; TV: tricuspid valve.

Upon initial evaluation at the RSSA Fetomaternal Unit on 28 August 2025, comprehensive fetomaternal ultrasonography demonstrated a singleton live intrauterine fetus in cephalic presentation with biometric parameters corresponding to 34–36 weeks of gestation and an estimated fetal weight of 3,415 g. The amniotic fluid index was within normal limits at 21.57 cm, and the placenta was positioned at the fundus and posterior uterine corpus with Grade III maturation. Fetal echocardiographic assessment revealed marked cardiomegaly with severe right atrial enlargement. Based on spatial chamber orientation and outflow tract angulation, the examining team initially raised suspicion of Double Outlet Right Ventricle

(DORV). Electronic fetal heart rate monitoring via non-stress test (CTG-1, Figure 1A) demonstrated a baseline fetal heart rate of 141 beats/min, normal short-term variability (STV 5–10 bpm), three spontaneous accelerations exceeding 15 beats/min lasting over 15 seconds, and complete absence of decelerations or uterine contractions, fulfilling ACOG and FIGO criteria for a reassuring Category I tracing. Given the complexity of the suspected cardiac anomaly and the absence of immediate fetal compromise, outpatient close surveillance was instituted, and an urgent joint multidisciplinary consultation with the Pediatric Cardiology Division was scheduled for definitive diagnostic ascertainment.

(A) Baseline Electronic Cardiogram (CTG / NST) at 34–36 Weeks (28/08/2025)  
 Reassuring Category I Tracing: Baseline FHR 141 bpm, STV 5–10 bpm, Accelerations >15 bpm for >15 s, No Decelerations



(B) Follow-up Electronic Cardiogram (CTG / NST) at 36–37 Weeks (11/09/2025)  
 Stable Category I Tracing: Baseline FHR 150 bpm, Robust STV/LTV 5–15 bpm, Spontaneous Accelerations, Absence of Fetal Distress



(C) Longitudinal Antenatal Maternal-Fetal Surveillance & Objective Category I Verification Matrix

| Surveillance Parameter           | Baseline NST (28 Aug 2025)             | Follow-Up NST (11 Sep 2025)            | Term Admission (18 Sep 2025)     | Clinical Assessment                      |
|----------------------------------|----------------------------------------|----------------------------------------|----------------------------------|------------------------------------------|
| Gestational Age & Presentation   | 34–36 Weeks / Cephalic                 | 36–37 Weeks / Cephalic                 | 38–39 Weeks / Cephalic           | Normal fetal somatic growth              |
| Baseline Fetal Heart Rate        | 141 beats/min (Normal)                 | 150 beats/min (Normal)                 | 147 beats/min (Normal)           | Preserved sinus pacemaker rhythm         |
| Heart Rate Variability (STV/LTV) | 5–10 bpm (Moderate)                    | 5–15 bpm (Moderate/Robust)             | 6–12 bpm (Moderate)              | Intact fetal autonomic innervation       |
| Fetal Heart Rate Accelerations   | Present ( $\geq 3$ in 20 min, >15 bpm) | Present ( $\geq 2$ in 20 min, >15 bpm) | Present (Reactive)               | Reassuring physiological autonomic drive |
| Pathological Decelerations       | Absent (No variable/late/prolonge)     | Absent (No variable/late/prolonge)     | Absent                           | Absence of uteroplacental insufficiency  |
| Uterine Contraction Activity     | Quiescent (0 contractions/10 min)      | Quiescent (0 contractions/10 min)      | Quiescent (Pre-labor)            | No premature labor induction             |
| AFI & Amniotic Fluid Status      | 21.57 cm (Normal AFI)                  | 19.8 cm (Normal AFI)                   | 18.5 cm (Normal AFI)             | Preserved fetal renal perfusion          |
| FIGO / ACOG CTG Classification   | Category I (Normal / Reassuring)       | Category I (Normal / Reassuring)       | Category I (Normal / Reassuring) | Permitted planned term delivery          |

**Figure 1. Antenatal electronic cardiogram (CTG / Non-Stress Test) tracings and longitudinal maternal-fetal surveillance matrix. (A) Baseline CTG tracing at 34–36 weeks of gestation (28/08/2025) demonstrating a baseline fetal heart rate of 141 beats/min, reassuring short-term variability (STV 5–10 bpm), and distinct physiological accelerations (>15 bpm lasting >15 s) fulfilling FIGO/ACOG Category I criteria. (B) Follow-up CTG tracing at 36–37 weeks of gestation (11/09/2025) demonstrating autonomic stability with baseline FHR of 150 bpm and robust variability (5–15 bpm). (C) Comprehensive surveillance matrix confirming absence of in utero fetal acidemia or heart block, permitting planned term delivery.**

On 11 September 2025 (36–37 weeks of gestation), repeat fetomaternal ultrasonography performed jointly with the Pediatric Cardiology team led to a definitive diagnostic revision: the initial suspicion of DORV was revoked and superseded by a diagnosis of suspected Ebstein's anomaly associated with multiple congenital anomalies. Fetal echocardiography (demonstrated in Figure 2A) revealed distinct apical

displacement of the septal and posterior tricuspid valve leaflets toward the right ventricular apex exceeding 8 mm/m<sup>2</sup> relative to the anterior mitral valve annulus insertion plane, significant atrialization of the inflow portion of the right ventricle, and severe right atrial volume overload. The detailed diagnostic differential matrix distinguishing this pathology from the initial regional suspicion of Double Outlet Right Ventricle is

comprehensively illustrated in Figure 2B. Repeat non-stress testing (demonstrated in Figure 1B) confirmed persistent autonomic stability, with a baseline heart rate of 150 beats/min, robust variability of 5–15 beats/min, positive accelerations, and absence of decelerations (Category I). A comprehensive multidisciplinary conference comprising maternal-fetal medicine specialists, pediatric cardiologists, neonatologists, and obstetric anesthesiologists was

convened. Given the complex nature of the cardiac lesion, the anticipated requirement for specialized neonatal intensive care, and the patient's explicit request for permanent contraception, elective lower segment cesarean section combined with immediate postpartum intrauterine device (IUD) insertion was scheduled for 19 September 2025 at 38–39 weeks of gestation.

(B) Diagnostic Differentiation: Initial Conotruncal Suspicion vs. Ebstein Anomaly

(A) Authentic Fetal Echocardiogram: Apical 4-Chamber View (34–36 Weeks)  
 Displays Severe Right Atrial Dilatation, Apical TV Displacement, aRV, & fRV



| Initial / Hemodynamic       | Initial Referral: DORV Suspicion    | Revised Diagnosis: Ebstein Anomaly                        |
|-----------------------------|-------------------------------------|-----------------------------------------------------------|
| Chamber Asymmetry & C       | Suspected RV dominance              | Profound right atrial enlargement & extensive             |
| Tricuspid Valve Insertion   | Assumed normal atrioventricular     | Marked apical displacement of septal/posterior            |
| Functional Right Ventricle  | Considered hypertrophied            | Diminutive, non-compliant chamber; impaired               |
| Great Artery Alignment      | Both great arteries suspected       | Concordant ventriculoarterial alignment; PA transposition |
| Color Doppler Regurgitation | Interventricular shunting           | Torrential systolic tricuspid regurgitation directed      |
| Interatrial Septum & Shunt  | Preserved foramen ovale             | Obligatory right-to-left shunt decompressor               |
| Translational Clinical Imp  | Risk of erroneous surgical approach | Enabled precise peripartum planning & airway              |

**Figure 2. Fetal echocardiographic ascertainment at 34–36 weeks of gestation (28/08/2025) and diagnostic revision from initial conotruncal suspicion. (A) High-resolution apical four-chamber fetal echocardiogram (GE Voluson E10) displaying marked right atrial (RA) dilatation, downward apical displacement of the tricuspid valve septal and posterior leaflets exceeding 8 mm/m<sup>2</sup>, an extensive atrialized right ventricle (aRV), a diminutive functional right ventricle (fRV), patent foramen ovale (FO), and left-sided chambers (LA and LV). (B) Detailed diagnostic differential matrix illustrating the spatial rotation pitfall that mimicked Double Outlet Right Ventricle (DORV) on initial routine obstetric screening.**

On 18 September 2025, the patient was admitted to Inpatient Ward 3 of RSSA for planned preoperative optimization. Clinical examination revealed a fully conscious, moderately ill-appearing woman with normal vital signs: blood pressure 118/78 mmHg, pulse rate 100 beats/min, respiratory rate 20 breaths/min, body temperature 36.5 °C, and oxygen saturation 98% breathing ambient air. Her height was 150 cm, current body weight was 66 kg (pre-pregnancy weight 51 kg), yielding a body mass index of 22.7 kg/m<sup>2</sup>. General physical examination showed pink conjunctivae, anicteric sclerae, normal vesicular breath sounds without wheezing or rhonchi, and regular cardiac rhythm with normal S1/S2 heart sounds without pathological murmurs. Obstetric examination demonstrated a fundal height of 33 cm, cephalic presentation, estimated fetal weight of 3,100 g, and fetal heart rate of 147 beats/min without uterine contractions or cervical dilation. Preoperative maternal laboratory investigations and fetal biophysical profiling are summarized in Table 2.

Hematological analysis demonstrated a hemoglobin of 10.8 g/dL, leukocyte count of 12,320 /μL, and platelet count of 387,000 /μL. Coagulation parameters were normal, with a prothrombin time of 15.0 seconds and activated partial thromboplastin time of 15.0 seconds. Metabolic and renal panels showed random blood glucose of 149 mg/dL, blood urea of 12.2 mg/dL, and serum creatinine of 0.55 mg/dL. Electrolyte panels confirmed serum sodium of 138 mmol/L, potassium of 3.31 mmol/L, chloride of 106 mmol/L, and serum albumin of 3.69 g/dL. Preoperative ultrasound confirmed persistent marked fetal cardiomegaly (cardiothoracic ratio 0.62, Figure 3B), a dilated main pulmonary artery trunk (Figure 3A), downward tricuspid valve displacement, and coexistent extracardiac anomalies including prominent bilateral scrotal hydrocele and external genital anomalies suggestive of hypospadias or phimosis (Figure 3C). Standard preoperative prophylaxis included intravenous cefazolin (2 g), ranitidine (50 mg), and metoclopramide (10 mg).

**Table 2. Maternal Preoperative Laboratory Investigations, Cardiotocographic Parameters, and Fetal Biophysical Profiling at Term Admission (18 September 2025).**

| Clinical Diagnostic Category                        | Parameter / Biomarker                        | Measured Value              | Standard Reference Interval                                                                                                                                                                                                       | Analytical Interpretation & Pathophysiological Significance                                                                                                                                          |
|-----------------------------------------------------|----------------------------------------------|-----------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Hematological Panel                                 | Hemoglobin (Hb)                              | 10.8 g/dL                   | 11.0 - 15.0 g/dL                                                                                                                                                                                                                  | Mild physiological gestational dilution; leukocytosis consistent with late third-trimester physiological stress; adequate platelet count ensuring safe spinal anesthesia and surgical hemostasis.    |
|                                                     | Leukocyte Count (WBC)                        | 12,320 /μL                  | 4,000 - 10,000 /μL                                                                                                                                                                                                                |                                                                                                                                                                                                      |
|                                                     | Thrombocyte Count (Platelets)                | 387,000 /μL                 | 150,000 - 450,000 /μL                                                                                                                                                                                                             |                                                                                                                                                                                                      |
|                                                     | Hematocrit (Hct)                             | 33.2 %                      | 35.0 - 47.0 %                                                                                                                                                                                                                     |                                                                                                                                                                                                      |
|                                                     |                                              |                             |                                                                                                                                                                                                                                   |                                                                                                                                                                                                      |
| Coagulation Profile                                 | Prothrombin Time (PT)                        | 15.0 seconds                | 11.0 - 15.0 seconds                                                                                                                                                                                                               | Normal extrinsic pathway activity; mildly shortened APTT reflecting typical physiological hypercoagulability of term pregnancy; absence of consumptive coagulopathy.                                 |
|                                                     | Activated Partial Thromboplastin Time (APTT) | 15.0 seconds                | 25.0 - 35.0 seconds                                                                                                                                                                                                               |                                                                                                                                                                                                      |
|                                                     |                                              |                             |                                                                                                                                                                                                                                   |                                                                                                                                                                                                      |
| Metabolic & Renal Profile                           | Random Blood Glucose (RBS)                   | 149 mg/dL                   | 70 - 140 mg/dL                                                                                                                                                                                                                    | Mild transient postprandial glycemic elevation without gestational diabetes criteria; preserved maternal renal filtration function and adequate intravascular volume status.                         |
|                                                     | Blood Urea                                   | 12.2 mg/dL                  | 10.0 - 50.0 mg/dL                                                                                                                                                                                                                 |                                                                                                                                                                                                      |
|                                                     | Serum Creatinine                             | 0.55 mg/dL                  | 0.50 - 0.90 mg/dL                                                                                                                                                                                                                 |                                                                                                                                                                                                      |
|                                                     |                                              |                             |                                                                                                                                                                                                                                   |                                                                                                                                                                                                      |
| Electrolytes & Liver Function                       | Serum Sodium (Na+)                           | 138 mmol/L                  | 135 - 145 mmol/L                                                                                                                                                                                                                  | Stable normonatremic electrolyte balance; borderline low potassium managed with dietary intake; preserved oncotic pressure maintaining intravascular stability and preventing maternal tissue edema. |
|                                                     | Serum Potassium (K+)                         | 3.31 mmol/L                 | 3.50 - 5.00 mmol/L                                                                                                                                                                                                                |                                                                                                                                                                                                      |
|                                                     | Serum Chloride (Cl-)                         | 106 mmol/L                  | 98 - 107 mmol/L                                                                                                                                                                                                                   |                                                                                                                                                                                                      |
|                                                     | Serum Albumin                                | 3.69 g/dL                   | 3.50 - 5.00 g/dL                                                                                                                                                                                                                  |                                                                                                                                                                                                      |
| Electronic Fetal Monitoring (CTG / Non-Stress Test) | Baseline Fetal Heart Rate                    | 150 beats/min               | Non-stress test demonstrated robust fetal autonomic cardiac regulation, intact sinoatrial nodal responsiveness, and absence of baseline bradyarrhythmias, tachyarrhythmias, or uteroplacental insufficiency.                      |                                                                                                                                                                                                      |
|                                                     | Baseline FHR Variability                     | 5 - 15 beats/min            |                                                                                                                                                                                                                                   |                                                                                                                                                                                                      |
|                                                     | Accelerations                                | Present (>2 in 20 min)      |                                                                                                                                                                                                                                   |                                                                                                                                                                                                      |
|                                                     | Decelerations                                | Absent                      |                                                                                                                                                                                                                                   |                                                                                                                                                                                                      |
|                                                     | Uterine Contractions                         | 0 contractions / 30 min     |                                                                                                                                                                                                                                   |                                                                                                                                                                                                      |
|                                                     | FIGO / ACOG Category                         | Category I (Normal)         |                                                                                                                                                                                                                                   |                                                                                                                                                                                                      |
| Fetomaternal Ultrasound Biometry & Fluid            | Biparietal Diameter (BPD)                    | 9.2 cm                      | Biometric dimensions corresponding to appropriate-for-gestational-age fetal growth (50th-75th percentile); upper-normal amniotic fluid volume (AFI 21.57 cm) ruling out overt polyhydramnios or oligohydramnios; mature placenta. |                                                                                                                                                                                                      |
|                                                     | Femur Length (FL)                            | 7.1 cm                      |                                                                                                                                                                                                                                   |                                                                                                                                                                                                      |
|                                                     | Abdominal Circumference (AC)                 | 33.8 cm                     |                                                                                                                                                                                                                                   |                                                                                                                                                                                                      |
|                                                     | Estimated Fetal Weight (EFW)                 | 3,415 g                     |                                                                                                                                                                                                                                   |                                                                                                                                                                                                      |
|                                                     | Amniotic Fluid Index (AFI)                   | 21.57 cm                    |                                                                                                                                                                                                                                   |                                                                                                                                                                                                      |
|                                                     | Placental Graded Maturation                  | Fundal-Posterior, Grade III |                                                                                                                                                                                                                                   |                                                                                                                                                                                                      |
|                                                     |                                              |                             |                                                                                                                                                                                                                                   |                                                                                                                                                                                                      |
|                                                     |                                              |                             |                                                                                                                                                                                                                                   |                                                                                                                                                                                                      |

ACOG: American College of Obstetricians and Gynecologists; AFI: amniotic fluid index; APTT: activated partial thromboplastin time; CTG: cardiotocography; FIGO: International Federation of Gynecology and Obstetrics; FHR: fetal heart rate; PT: prothrombin time; RBS: random blood sugar; WBC: white blood cells.

(A) Fetal Echocardiography: Pulmonary Artery Dilatation (38–39 Weeks)  
 Right Ventricular Outflow Tract View Demonstrating Compensatory Dilated PA Trunk



(B) Fetal Ultrasonography: Severe Cardiomegaly (38–39 Weeks)  
 Transverse Thoracic Scan Demonstrating Cardiothoracic Ratio (CTR) of 0.62



(C) Fetal Perineal Ultrasonography: Extracardiac Anomalies  
 Demonstrates Prominent Bilateral Testicular Hydrocele & Atypical Penile Contour



(D) Embryological Synchronization of Coexistent Malformations  
 Shared Organogenesis Vulnerability Window (Gestational Weeks 6–14)

SHARED EMBRYOLOGICAL TIMELINE & SIGNALING PATHWAYS:

1. Tricuspid Valve Delamination Arrest (Gestational Weeks 6–10):
  - Physiologic Mechanism: BMP-2/4, Notch, and canonical Wnt/ $\beta$ -catenin regulate myocardial cavitation and enzymatic cleavage of valve tissue.
  - Pathological Insult: Delamination failure leaves septal/posterior leaflets fused to RV trabeculae, creating an atrialized RV (aRV) and tricuspid regurgitation.
2. External Genitalia Differentiation & Processus Vaginalis (Weeks 8–14):
  - Urethral Groove Fusion: Androgen receptor signaling and Sonic hedgehog (Shh) orchestrate midline closure of urethral folds forming the penile urethra.
  - Processus Vaginalis Patency: Incomplete obliteration of the peritoneal evagination permits peritoneal fluid accumulation, precipitating bilateral scrotal hydrocele.
3. Gene-Environment Interaction & Maternal Exposure Hypothesis:
  - Maternal history negative for pharmacological teratogens (lithium carbonate).
  - Chronic secondary exposure to arterial vehicle exhaust & passive cigarette smoke.
  - Combustion-derived polycyclic aromatic hydrocarbons & PM<sub>2.5</sub> induce systemic oxidative stress and epigenetic methylation shifts during overlapping weeks 6–10.
4. Clinical Translation & Management Implication:
  - Benign, self-limiting nature of hydrocele confirmed by spontaneous resolution at 3 mo.
  - Averted unnecessary invasive urological intervention during fragile neonatal period.

**Figure 3. Advanced multidisciplinary fetomaternal ultrasonography at 38–39 weeks of gestation (18/09/2025) and characterization of coexistent extracardiac malformations. (A) Oblique right ventricular outflow tract sweep demonstrating compensatory dilatation of the main pulmonary artery trunk (A. Pulmonalis). (B) Transverse thoracic scan confirming persistent severe cardiomegaly with a cardiothoracic ratio (CTR) of 0.62. (C) High-resolution targeted coronal view of the fetal perineum demonstrating bilateral anechoic fluid collections within the scrotal sac consistent with bilateral testicular hydrocele, alongside an atypical penile contour suggestive of hypospadias vs. phimosis. (D) Embryological developmental synthesis card highlighting shared organogenesis vulnerability during gestational weeks 6 to 14.**

On 19 September 2025 at 08:25 AM, elective lower segment cesarean section was successfully performed under regional spinal anesthesia. A vigorous live male infant was delivered weighing 3,050 g, measuring 49 cm in length, with an estimated gestational age of 38–39 weeks, and Apgar scores of 5 at one minute and 7 at five minutes. Amniotic fluid was clear upon hysterotomy. Immediate postpartum insertion of a copper T-380A intrauterine device at the uterine fundus was accomplished prior to two-layer hysterorrhaphy. Total intraoperative blood loss was approximately 300 mL, and the maternal surgical procedure was completed without complication. Postpartum maternal recovery was uneventful, with satisfactory Pfannenstiel wound healing, gradual mobilization, and discharge on postpartum day 3. Outpatient surgical check on postpartum day 11 (30 September 2025) confirmed complete incision epithelialization and normal uterine involution.

The neonate's serial hemodynamic, echocardiographic, and oxygenation trajectory from delivery through 3-month follow-up is chronologically presented in Table 3 and illustrated in Figure 4. Immediately following delivery, the neonate exhibited mild central cyanosis with an initial peripheral oxygen saturation (SpO<sub>2</sub>) of 85% on room air, respiratory rate of 55 breaths/min, heart rate of 135 beats/min, warm peripheral extremities with a capillary refill time under 2 seconds, and an audible systolic murmur over the lower left sternal border. The infant was transferred immediately to the level III NICU. At two hours of life (10:21 AM), urgent bedside neonatal transthoracic echocardiography performed by the Pediatric Cardiology team definitively established the diagnosis of Ebstein's anomaly (Carpentier Type B). Key echocardiographic findings included marked downward displacement of the septal and posterior tricuspid leaflets, a large atrialized right ventricle, and moderate-to-severe tricuspid regurgitation. Doppler

interrogation revealed a maximum tricuspid regurgitant velocity (Vmax) of 4.4 m/s, yielding a peak systolic pressure gradient of 78 mmHg, indicative of elevated right ventricular systolic pressure and a high probability of persistent pulmonary hypertension of the newborn (PPHN). Crucially, an atrial septal defect (ASD) measuring 3–4 mm was identified with obligatory right-to-left shunting, serving as an essential decompression valve for the hypertensive right atrium. Furthermore, a patent ductus arteriosus (PDA) measuring 3 mm was visualized with continuous left-

to-right shunting, demonstrating that systemic arterial pressure in the descending aorta was actively driving blood flow into the pulmonary trunk to maintain adequate pulmonary perfusion. Left ventricular systolic function was preserved with an ejection fraction (LVEF) of 78%, and right ventricular longitudinal excursion (TAPSE) was 1.0 cm. Neonatal blood analysis showed hemoglobin 13.2 g/dL, leukocyte count 12,000 /μL, platelet count 378,000 /μL, C-reactive protein 0.1 mg/L, and I/T ratio 0.05.

**Table 3. Serial Neonatal Hemodynamic, Echocardiographic, and Oxygenation Trajectory from Delivery to 3-Month Follow-Up.**

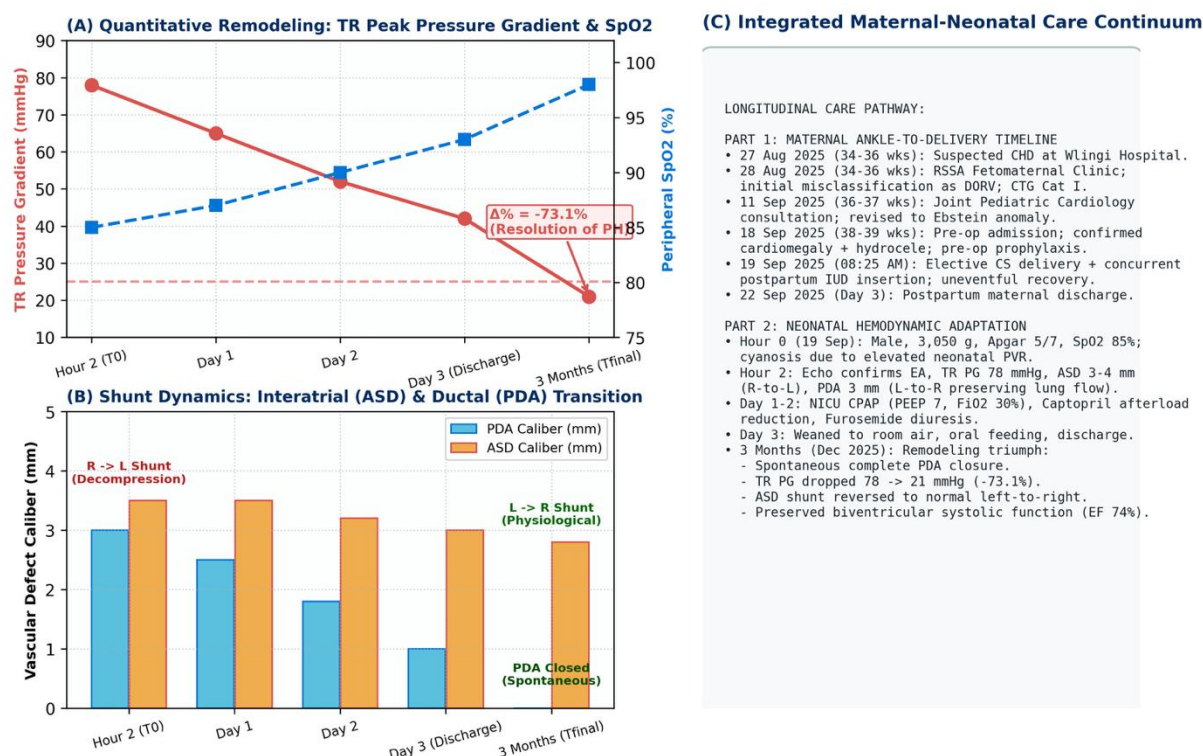
| Timepoint & Clinical Stage                                           | Vital Signs & Gas Exchange                                                                                                       | Key Echocardiographic Biomarkers & Hemodynamics                                                                                                                                                                                              | Respiratory & Pharmacological Management                                                                                                                                                              | Hemodynamic State & Clinical Trajectory                                                                                                                                             |
|----------------------------------------------------------------------|----------------------------------------------------------------------------------------------------------------------------------|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Hour 0 - 2<br>(19 Sep 2025, 08:25)<br>Delivery & NICU Admission      | HR: 135 bpm<br>RR: 55 breaths/min<br>SpO2: 85% (room air)<br>Temp: 36.7 °C<br>CRT: <2 seconds<br>Apgar: 5 (1 min) / 7 (5 min)    | Downward displacement of septal/posterior tricuspid valve leaflets; marked right atrial enlargement; functional right ventricular hypoplasia.                                                                                                | Immediate airway suctioning, thermal stabilization; transfer to level III NICU; continuous pulse oximetry monitoring.                                                                                 | Mild central cyanosis at birth (SpO2 85%) due to obligatory right-to-left interatrial shunting in the presence of elevated neonatal PVR.                                            |
| Hour 2 - 8<br>(19 Sep 2025, 10:21)<br>Initial Echocardiography       | HR: 135 bpm<br>RR: 55 breaths/min<br>SpO2: 85% - 88%<br>BP: 64/38 mmHg<br>CRT: <2 seconds<br>Afebrile, active tone               | Ebstein anomaly confirmed: moderate-to-severe TR; TR Vmax 4.4 m/s, peak PG 78 mmHg (high probability pulmonary hypertension); ASD 3–4 mm (right-to-left shunt); PDA 3 mm (left-to-right shunt); LVEF 78%; TAPSE 1.0 cm.                      | CPAP commenced (PEEP 7 cmH2O, FiO2 30%); NPO; IV maintenance fluids (60 mL/kg/day); IV Ampicillin-Sulbactam + Chloramphenicol (suspected neonatal pneumonia); Paracetamol.                            | Transitional circulation stabilized: 3 mm PDA left-to-right shunt preserves pulmonary perfusion; ASD decompresses high right atrial pressure; preserved LV ejection fraction (78%). |
| Postnatal Day 1<br>(20 Sep 2025, 10:48)<br>Therapeutic Stabilization | HR: 135 bpm<br>RR: 52 breaths/min<br>SpO2: 85% - 89% (on CPAP)<br>Temp: 36.7 °C<br>CRT: <2 seconds<br>Target SpO2: 75% - 85%     | Persistent right-to-left interatrial communication maintaining systemic cardiac output; stable ductal caliber with continuous left-to-right flow; no retrograde pulmonary flow.                                                              | CPAP PEEP 7 cmH2O, FiO2 30%; fluid advanced to 80 mL/kg/day; oral Captopril initiated (0.1 mg/kg/dose q8h) to reduce afterload; oral Furosemide (1 mg/kg/dose q12h); platelet transfusion (TC 60 mL). | Hemodynamic equilibrium achieved: avoiding excessive oxygenation prevents precipitous ductal constriction while afterload reduction supports right ventricular emptying.            |
| Postnatal Day 2<br>(21 Sep 2025, 08:44)<br>Weaning Phase             | HR: 130 bpm<br>RR: 48 breaths/min<br>SpO2: 88% - 91% (low-flow nasal cannula)<br>Temp: 36.8 °C<br>CRT: <2 seconds                | Gradual decrease in estimated right ventricular systolic pressure; stable right-sided chamber dimensions; no pericardial effusion; intact systemic perfusion.                                                                                | Successful wean from CPAP to low-flow nasal cannula (0.5 L/min, room air); trophic oral feeding with expressed breast milk initiated; continuation of Captopril and Furosemide.                       | Clinical transition from respiratory support dependence to independent spontaneous breathing; physiological PVR decline underway.                                                   |
| Postnatal Day 3<br>(22 Sep 2025)<br>Discharge Assessment             | HR: 128 bpm<br>RR: 42 breaths/min<br>SpO2: 92% - 94% (room air)<br>Temp: 36.6 °C<br>Weight: 3,020 g<br>Vigorous suckling         | Adequate antegrade forward pulmonary flow; moderate tricuspid regurgitation; closing ductus arteriosus; preserved biventricular systolic performance.                                                                                        | Discharge on oral Captopril (0.15 mg/kg/dose q8h) and oral Furosemide (1 mg/kg/dose q12h); mother instructed on signs of cardiac decompensation; outpatient cardiology follow-up booked.              | Successful non-surgical neonatal discharge; avoidance of emergency neonatal cone repair or Blalock-Taussig shunt.                                                                   |
| 3-Month Follow-Up<br>(15 Dec 2025)<br>Longitudinal Remodeling        | HR: 122 bpm<br>RR: 34 breaths/min<br>SpO2: 98% (room air)<br>Weight: 5,850 g (50th percentile)<br>Normal psychomotor development | Remarkable structural remodeling: TR peak PG plummeted to 21 mmHg ( $\Delta\%$ = -73.1%, resolution of pulmonary hypertension); spontaneous complete PDA closure; ASD shunt reversed to physiological left-to-right; LVEF 74%; TAPSE 1.2 cm. | Gradual taper of oral Furosemide; continuation of low-dose Captopril maintenance; pediatric urology consultation for bilateral hydrocele resolution and hypospadias surveillance.                     | Definitive hemodynamic adaptation: normalization of pulmonary vascular resistance transformed severe neonatal disease into stable, biventricular pediatric physiology.              |

ASD: atrial septal defect; BP: blood pressure; CPAP: continuous positive airway pressure; CRT: capillary refill time; FiO2: fraction of inspired oxygen; HR: heart rate; LVEF: left ventricular ejection fraction; NPO: nil per os; PDA: patent ductus arteriosus; PEEP: positive end-expiratory pressure; PG: pressure gradient; PVR: pulmonary vascular resistance; RR: respiratory rate; SpO2: peripheral capillary oxygen saturation; TAPSE: tricuspid annular plane systolic excursion; TC: thrombocyte concentrate; TR: tricuspid regurgitation; Vmax: maximum velocity;  $\Delta\%$ : relative percentage change.

Respiratory support was initiated utilizing continuous positive airway pressure (CPAP) via binasal prongs with a positive end-expiratory pressure (PEEP) of 7 cmH<sub>2</sub>O and an inspired oxygen fraction (FiO<sub>2</sub>) of 30%, which maintained SpO<sub>2</sub> within the targeted transitional range of 85% to 89%. Intravenous fluid therapy was administered at 60 mL/kg/day, transitioning to 80 mL/kg/day on postnatal day 1, alongside empirical intravenous antibiotic therapy (ampicillin-sulbactam and chloramphenicol) for suspected neonatal pneumonia. To reduce right and left ventricular afterload and promote antegrade cardiac output, oral captopril was initiated at 0.1 mg/kg/dose every 8 hours, combined with oral furosemide at 1 mg/kg/dose every 12 hours for volume decongestion. Because pulmonary blood flow was robustly maintained via the left-to-right ductal shunt without anatomical pulmonary atresia, prostaglandin E1 infusion was deliberately withheld, avoiding unnecessary pharmacological vasodilation that could destabilize systemic perfusion. Over postnatal days 1 and 2, the infant demonstrated progressive clinical stabilization. Respiratory support was successfully weaned from CPAP to low-flow nasal cannula (0.5 L/min on room air) and subsequently to ambient air, with SpO<sub>2</sub> rising to 92%–94%. Enteral feeding with expressed breast milk was advanced without feeding intolerance. The infant was discharged home on postnatal day 3 in stable hemodynamic condition on maintenance oral

captopril (0.15 mg/kg/dose every 8 hours) and furosemide (1 mg/kg/dose every 12 hours).

At the 3-month comprehensive outpatient follow-up evaluation on 15 December 2025, the infant demonstrated thriving growth, weighing 5,850 g (50th percentile) with normal neuromotor milestones, no cyanosis, and SpO<sub>2</sub> of 98% on room air. Repeat transthoracic echocardiography documented remarkable structural and hemodynamic remodeling (tracked longitudinally in Figure 4A): the tricuspid regurgitant peak velocity had decreased markedly, and the peak systolic pressure gradient had plummeted from the initial 78 mmHg to 21 mmHg—representing an extraordinary relative reduction of 73.1% ( $\Delta\% = -73.1\%$ ) and confirming complete resolution of neonatal pulmonary hypertension. Furthermore, the patent ductus arteriosus had closed completely and spontaneously, while the interatrial communication had reversed its shunt directionality to a physiological left-to-right pattern across a small 2.8 mm secundum ASD (depicted in Figure 4B). Both left and right ventricular systolic performance remained fully preserved (LVEF 74%, TAPSE 1.2 cm), with the integrated maternal-neonatal clinical care continuum flowchart detailed in Figure 4C. Clinical examination by pediatric surgery confirmed spontaneous, complete resolution of the bilateral scrotal hydrocele, with conservative monitoring planned for the mild distal hypospadias.



**Figure 4. Longitudinal quantitative hemodynamic trajectory and serial biventricular remodeling from acute neonatal presentation to 3-month follow-up. (A) Time-course trajectory of peak tricuspid regurgitant (TR) systolic pressure gradient (mmHg, red curve) and peripheral capillary oxygen saturation (SpO<sub>2</sub> %, blue curve), highlighting a 73.1% reduction in TR gradient from 78 mmHg to 21 mmHg ( $\Delta\% = -73.1\%$ ), confirming complete resolution of neonatal pulmonary hypertension. (B) Caliber and shunt directionality dynamics of the patent ductus arteriosus (PDA, cyan bars) and atrial septal defect (ASD, orange bars), displaying spontaneous ductal closure and physiological shunt reversal from right-to-left to left-to-right. (C) Integrated maternal-neonatal care continuum flowchart.**

Objective diagnostic and prognostic risk stratification for the patient is detailed in Table 4. The calculated Celermajer Index at end-diastole was 0.88, categorizing the infant as Grade 2 (intermediate risk, predicted perinatal mortality ~10%), which accurately aligned with the infant's favorable survival under medical therapy and ruled out the catastrophic mortality (>90%) associated with Grade 4 disease (Celermajer Index  $\geq 1.50$ ). The fetal Severity Assessment Score (SAS) was 4 points (intermediate risk), reflecting marked cardiomegaly and tricuspid regurgitation but tempered by critical protective features: complete absence of hydrops fetalis, absence of pulmonary regurgitation, and preserved antegrade pulmonary trunk connection without a circular shunt. The Fetal Cardiovascular Profile Score (CVPS) was 8 out of 10 (deductions of 1 point for cardiothoracic ratio >0.50

and 1 point for holosystolic TR), indicating stable in utero compensation that justified planned term delivery. Anatomically, the lesion conformed to Carpentier Type B, providing sufficient functional right ventricular myocardial mass and an adequately mobile anterior leaflet to support forward pulmonary flow once pulmonary vascular resistance normalized postnatally. Table 5 provides a global comparative benchmarking of the present study against leading international prenatally diagnosed Ebstein's anomaly cohorts published between 2021 and 2026, highlighting the convergent necessity of interatrial decompression, the decisive protective role of non-circular shunt physiology, and the unique demonstration of spontaneous ductal-dependent resolution achieved without early neonatal cardiac surgery in a Southeast Asian academic medical center.

**Table 4. Objective Diagnostic Scoring and Prognostic Risk Stratification Matrix for the Present Case.**

| Prognostic Scoring System                                 | Parameters Evaluated & Methodological Basis                                                                                                                                                                                                         | Present Case Calculated Value                                                                                                                        | Standard Stratification Cut-offs                                                                                                                                                    | Predicted Mortality & Clinical Trajectory Correlation                                                                                                                                                            |
|-----------------------------------------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Celermajer Index (Great Ormond Street Score)              | Ratio of the combined area of the right atrium and atrialized right ventricle to the combined area of the functional right ventricle, left atrium, and left ventricle at end-diastole: $(RA + aRV) / (fRV + LA + LV)$ .                             | Score: 0.88 (Grade 2)                                                                                                                                | Grade 1: < 0.50 (Mortality ~8%)<br>Grade 2: 0.50 - 0.99 (Mortality ~10%)<br>Grade 3: 1.00 - 1.49 (Mortality ~44%)<br>Grade 4: $\geq 1.50$ (Mortality >90%)                          | Accurately predicted favorable neonatal survival with medical management; avoided the catastrophic mortality (>90%) associated with Grade 4 extreme cardiomegaly.                                                |
| Severity Assessment Score (SAS) for Fetal Ebstein Anomaly | Multivariate score incorporating cardiothoracic ratio (CTR >0.55), tricuspid displacement index, presence of pulmonary regurgitation (circular shunt), retrograde ductal flow, and LV compression.                                                  | Score: 4 points (Intermediate Risk)                                                                                                                  | Low Risk: 0 - 2 points<br>Intermediate Risk: 3 - 5 points<br>High Risk: 6 - 8 points                                                                                                | Intermediate risk reflected marked cardiomegaly and TR but with critical protective features: absence of in utero hydrops, absence of circular shunt (no pulmonary regurgitation), and antegrade pulmonary flow. |
| Cardiovascular Profile Score (CVPS - Huhta / AHA)         | Comprehensive 10-point composite score assessing 5 categories (2 points each): hydrops, cardiomegaly (CTR), cardiac function/effusion, arterial Doppler, and venous Doppler.                                                                        | Score: 8 / 10 (Deductions: -1 for CTR >0.50, -1 for mild holosystolic TR)                                                                            | Normal / Low Risk: 9 - 10 points<br>Moderate Risk: 7 - 8 points<br>High Perinatal Demise: $\leq 6$ points                                                                           | A CVPS of 8/10 at 36–38 weeks signaled stable in utero compensation, ruling out immediate fetal distress and permitting planned term delivery rather than emergency preterm intervention.                        |
| Carpentier Anatomical Classification of Ebstein Anomaly   | Morphological classification based on leaflet mobility and degree of RV atrialization: Type A (mild), Type B (large atrialized component, mobile anterior leaflet), Type C (severely restricted anterior leaflet), Type D (sac-like atrialized RV). | Classification: Type B (Moderate atrialization, mobile sail-like anterior leaflet)                                                                   | Type A: Excellent reparability<br>Type B: Favorable for biventricular cone repair if needed<br>Type C: Complex repair required<br>Type D: Univentricular palliation often necessary | Type B anatomy provided sufficient functional RV volume and an adequately mobile anterior leaflet to sustain forward pulmonary blood flow once pulmonary vascular resistance declined postnatally.               |
| ISUOG 2023 Cardiac Screening Framework                    | Comprehensive 5-level examination protocol: situs, 4-chamber view, LV outflow tract, RV outflow tract, and 3-vessel-trachea (3VT) view with color Doppler.                                                                                          | Four-chamber asymmetry, apical TV displacement, and dilated pulmonary artery correctly identified on tertiary referral; missed on primary screening. | Basic 4-chamber view detection sensitivity: ~50–60%; Enhanced outflow tract + 3VT view sensitivity: >85–90%.                                                                        | Underlines the necessity of mandatory outflow tract and color Doppler evaluation in routine second-trimester screening to eliminate late third-trimester diagnostic delays.                                      |

AHA: American Heart Association; aRV: atrialized right ventricle; CTR: cardiothoracic ratio; CVPS: cardiovascular profile score; fRV: functional right ventricle; ISUOG: International Society of Ultrasound in Obstetrics and Gynecology; LA: left atrium; LV: left ventricle; RA: right atrium; SAS: severity assessment score; TR: tricuspid regurgitation; TV: tricuspid valve.

**Table 5. Global Comparative Literature Benchmarking Matrix of Prenatally Diagnosed Ebstein's Anomaly Cohorts (2021–2026).**

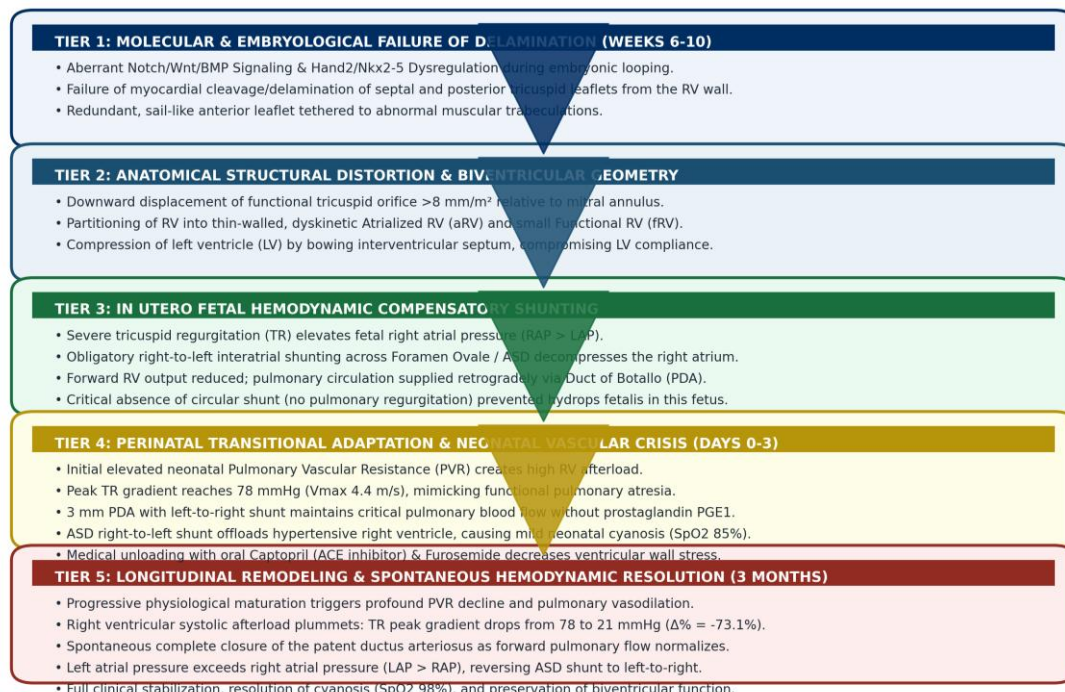
| Study Cohort & Year<br>(Lead Author / Country)                                             | Sample Size (N) &<br>Design Framework                      | Mean Gestational Age at Diagnosis (Weeks)                 | Associated Lesions & Circular Shunt Prevalence                                                                                                      | Neonatal Intervention & Perinatal Mortality Rates                                                                                          | Convergent Findings & Divergent Novelty of Present Study                                                                                                                                                                                          |
|--------------------------------------------------------------------------------------------|------------------------------------------------------------|-----------------------------------------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Kumar TKS et al. (2021)<br>India<br>doi:10.1007/s12055-020-00942-z                         | N = 48<br>Retrospective Tertiary Cohort (Neonates with EA) | Postnatal presentation (82%)<br>Prenatal diagnosis in 18% | ASD/PFO in 88%; RVOTO/pulmonary atresia in 32%; circular shunt in 14%.                                                                              | Surgical intervention in 45% (Starnes procedure, cone repair); early neonatal mortality 28%.                                               | Convergence: ASD serves as essential right-atrial decompression valve. Divergence: Present study demonstrated spontaneous clinical resolution without neonatal surgery, avoiding early operative mortality.                                       |
| Stepien A et al. / AATS (2024)<br>USA / International<br>doi:10.1016/j.jtcvs.2024.04.018   | Consensus Document<br>AATS Expert Consensus Guidelines     | Stratified prenatal (18–24 wks) vs late (>32 wks)         | Associated ASD >80%; circular shunt flagged as absolute high-mortality predictor (>80%).                                                            | Recommends planned delivery in level IV centers with ECMO; deferred surgery if forward flow exists.                                        | Convergence: Fully validates RSSA multidisciplinary protocol of term delivery, initial CPAP, and deferred surgical intervention. Divergence: Documents successful medical management in Southeast Asian academic center.                          |
| Wieczorek A et al. (2022)<br>Poland<br>doi:10.3390/jcm11133684                             | N = 54 fetuses<br>Prospective Multicenter Fetal Registry   | 24.2 ± 4.1 weeks (Second trimester)                       | Cardiomegaly (CTR >0.60) in 78%; circular shunt in 22%; pulmonary atresia in 18%.                                                                   | Perinatal mortality 44.4%; circular shunt mortality 91.6%; hydrops fetalis mortality 100%.                                                 | Convergence: Confirms that absence of circular shunt is the primary driver of neonatal survival. Divergence: Present study details late third-trimester ascertainment (34–36 wks) following initial DORV misdiagnosis.                            |
| McElhinney DB et al. (2021)<br>USA (Multi-institutional)<br>doi:10.1016/j.jacc.2021.02.039 | N = 124 neonates<br>Contemporary Multicenter Registry      | Prenatally diagnosed in 42%<br>Postnatal diagnosis in 58% | Secundum ASD in 91%; ductal-dependent pulmonary blood flow in 52%.                                                                                  | Biventricular circulation achieved in 68%; univentricular palliation in 22%; mortality 16%.                                                | Convergence: Demonstrates that neonates with preserved functional RV and mobile anterior leaflet achieve stable biventricular physiology. Divergence: Confirms spontaneous PDA closure with concurrent TR gradient drop (-73.1%).                 |
| Present Study (2026)<br>RSSA Malang, Indonesia<br>doi:10.37275/bsm.v10i8.1189              | N = 1 (In-depth Observational Analytic Longitudinal Study) | 34–36 weeks (Delayed referral from regional hospital)     | Downwards TV displacement, moderate TR, Secundum ASD (3–4 mm), PDA (3 mm), bilateral testicular hydrocele, hypospadias/phimosis; no circular shunt. | Zero operative neonatal mortality; managed conservatively with CPAP, Captopril, Furosemide; discharged Day 3; full remodeling at 3 months. | Core Novelty: Documents late prenatal diagnostic conversion (DORV to EA); serial hemodynamic remodeling showing TR PG drop from 78 to 21 mmHg; novel coexistence of EA with urogenital anomalies; tertiary protocol success in low-resource LMIC. |

AATS: American Association for Thoracic Surgery; ASD: atrial septal defect; CTR: cardiothoracic ratio; DORV: double outlet right ventricle; EA: Ebstein's anomaly; ECMO: extracorporeal membrane oxygenation; LMIC: low- and middle-income country; PFO: patent foramen ovale; PG: pressure gradient; PVR: pulmonary vascular resistance; RSSA: Rumah Sakit Umum Daerah Dr. Saiful Anwar; RV: right ventricle; RVOTO: right ventricular outflow tract obstruction; TR: tricuspid regurgitation; TV: tricuspid valve.

## 4. Discussion

This observational analytic study provides a rigorous, longitudinal treatise on the complex pathophysiology, prenatal diagnostic dilemmas, and transitional hemodynamic remodeling of Ebstein's anomaly diagnosed in the late third trimester. The overarching clinical trajectory of this patient—progressing from a delayed, misclassified prenatal referral to successful

non-surgical neonatal stabilization and subsequent dramatic hemodynamic remodeling at 3 months—offers several profound translational insights into congenital right ventricular malformations and perinatal care delivery in resource-variable settings. The multi-tier molecular, embryological, anatomical, and transitional cascade governing this clinical evolution is integrated conceptually in Figure 5.



**Figure 5. Comprehensive multi-tier molecular, embryological, anatomical, and transitional pathophysiological cascade of Ebstein's anomaly and perinatal circulatory adaptation. Delineates the sequential progression from Tier 1 (BMP/Notch/Wnt delamination failure at weeks 6–10), through Tier 2 (anatomical partitioning into atrialized and functional right ventricle), Tier 3 (fetal right-to-left interatrial compensatory shunting), Tier 4 (acute neonatal transitional crisis with high PVR, functional pulmonary atresia, and ductal left-to-right support), to Tier 5 (longitudinal remodeling with 73.1% plunge in TR gradient, spontaneous PDA closure, and shunt reversal).**

At the fundamental biological tier, Ebstein's anomaly represents a primary disorder of embryological delamination. During normal cardiogenesis, delamination is tightly regulated by complex spatiotemporal signaling pathways involving bone morphogenetic proteins (BMP-2 and BMP-4), the Notch signaling cascade, and canonical Wnt/ $\beta$ -catenin pathways, which orchestrate the progressive fenestration and separation of the inner myocardial mantle into discrete atrioventricular valve leaflets.<sup>1,2</sup> Disruption of this developmental genetic network—or adverse epigenetic modulation triggered by environmental exposures—arrests the cleavage planes, leaving the septal and posterior leaflets congenitally fused to the right ventricular trabeculae.<sup>2,6</sup> In our patient, maternal history was negative for pharmacological teratogens such as lithium carbonate, which has historically been linked to tricuspid valve delamination arrest via inositol depletion and glycogen synthase kinase-3 $\beta$  (GSK-3 $\beta$ ) inhibition.<sup>27</sup> However, the patient experienced chronic, heavy passive exposure to environmental pollutants, including vehicle exhaust particulate matter and tobacco combustion products.

Emerging epidemiological and experimental evidence suggests that maternal inhalation of combustion-derived polycyclic aromatic hydrocarbons and fine particulate matter (PM<sub>2.5</sub>) induces systemic endothelial oxidative stress, placental hypoxia, and aberrant DNA methylation in embryonic cardiac progenitor cells, potentially serving as an environmental cofactor in disrupted cardiac looping and valve delamination.<sup>21,25</sup> The anatomical phenotype in this infant conformed to Carpentier Type B, characterized by moderate apical displacement of the septal leaflet, significant atrialization of the inflow right ventricle, and a large, redundant, but mobile anterior leaflet.<sup>7</sup> This specific morphological substrate was decisive in determining the infant's favorable neonatal trajectory. In severe Carpentier Type C and Type D variants, the anterior leaflet is severely tethered to the ventricular free wall by dense, anomalous muscular cords, which not only prevents systolic coaptation but also physically obstructs the right ventricular outflow tract, virtually precluding effective antegrade stroke volume.<sup>7,8</sup> In contrast, the preserved mobility of the sail-like anterior

leaflet in Type B anatomy allows it to act as a functional flap-valve: while incompetent during high-afterload conditions, it can achieve partial coaptation once afterload diminishes, thereby directing forward stroke volume into the pulmonary trunk.<sup>7,10</sup>

The biophysical hemodynamics of the neonatal transitional circulation in Ebstein's anomaly represent one of the most perilous frontiers in pediatric cardiology, governed by the dynamic interplay between pulmonary vascular resistance, right ventricular compliance, and shunting pathways.<sup>18,28</sup> In the healthy neonate, pulmonary vascular resistance plummets precipitously within minutes of birth as lung inflation, mechanical alveolar expansion, and systemic oxygenation trigger rapid release of endogenous endothelial nitric oxide and prostacyclin, causing pulmonary arteriolar vasodilation.<sup>28,29</sup> In the neonate with Ebstein's anomaly, however, this transition is profoundly perturbed. The diminutive, highly non-compliant functional right ventricle is confronted by a normal, yet transiently elevated neonatal PVR. In our patient, this generated severe right ventricular afterload, elevating right ventricular systolic pressure and driving the tricuspid regurgitant velocity to 4.4 m/s, which yielded an exceptionally high peak pressure gradient of 78 mmHg during initial echocardiography at two hours of life. This degree of tricuspid hypertension frequently leads to functional pulmonary atresia, wherein the impaired right ventricle cannot generate sufficient systolic pressure to overcome pulmonary arterial closing pressure, causing forward flow across the pulmonary valve to cease completely.<sup>1,22</sup>

Under these high-afterload conditions, the survival of the neonate hinged entirely on the geometry and hemodynamics of the two communicating fetal shunts: the patent ductus arteriosus (PDA) and the atrial septal defect (ASD). The 3 mm PDA functioned as a critical life-sustaining conduit by delivering systemic arterial blood from the high-pressure descending aorta into the low-pressure pulmonary trunk via a left-to-right shunt, ensuring adequate pulmonary perfusion and alveolar gas exchange despite compromised right ventricular forward output.<sup>13,28</sup> Concurrently, the 3–4 mm ASD served as an indispensable decompression valve for the severely hypertensive, volume-overloaded right atrium. By permitting unrestricted right-to-left shunting of desaturated systemic venous return into the low-pressure left atrium, the ASD prevented catastrophic systemic venous congestion, hepatic capsular distension, and low-output circulatory collapse, while maintaining left ventricular filling and systemic cardiac output at the cost of mild, well-tolerated arterial desaturation (SpO<sub>2</sub> 85%).<sup>1,13</sup> This precise hemodynamic balance explains why the neonate maintained clinical stability without requiring emergency mechanical ventilation or inotropic support.

Crucially, the absence of a circular shunt was the primary biological determinant that averted early neonatal demise in this infant. In international registries of prenatally diagnosed Ebstein's anomaly, the presence of a circular shunt—defined as retrograde ductal flow combined with free pulmonary valve regurgitation—is associated with an overwhelming perinatal mortality rate exceeding 80% to 92%.<sup>15,18</sup> In our patient, detailed color Doppler echocardiography confirmed that the pulmonary valve was anatomically intact and competent, preventing retrograde regurgitation into the functional right ventricle. Consequently, blood shunted left-to-right across the ductus arteriosus was successfully directed into the pulmonary microvascular bed rather than recycling futilely through the heart. This critical physiological distinction allowed our clinical team to deliberately avoid intravenous prostaglandin E1 (PGE1) infusion. While PGE1 is standard therapy in ductal-dependent congenital heart lesions, its routine use in neonates with Ebstein's anomaly without anatomical pulmonary atresia can be deleterious: by maintaining ductal patency while simultaneously lowering systemic vascular resistance, PGE1 can worsen pulmonary overcirculation, promote retrograde ductal stealing from the systemic circulation, and exacerbate right ventricular volume overload.<sup>1,13,18</sup> Instead, our strategy focused on non-invasive respiratory support with CPAP (PEEP 7 cmH<sub>2</sub>O, FiO<sub>2</sub> 30%) to maintain functional residual capacity without alveolar hyperdistension, paired with pharmacologically targeted afterload reduction using oral captopril and gentle diuresis with furosemide.<sup>30,31</sup> Angiotensin-converting enzyme (ACE) inhibition with captopril effectively lowered systemic and pulmonary vascular tone, reducing right and left ventricular wall stress and promoting antegrade ejection, which facilitated rapid weaning from CPAP to ambient room air by postnatal day 2.<sup>30,31</sup>

The most striking longitudinal finding in this study was the extraordinary hemodynamic remodeling documented at the 3-month follow-up evaluation. Over the first several weeks of life, the physiological involution of the pulmonary arteriolar medial smooth muscle layer triggered a progressive, permanent decline in pulmonary vascular resistance.<sup>28,29</sup> As right ventricular afterload dissipated, the functional right ventricle was able to eject an increasingly large forward stroke volume through the pulmonary valve. This progressive unloading of the right heart culminated in a dramatic 73.1% plunge in the tricuspid regurgitant peak pressure gradient—dropping from an initial hypertensive value of 78 mmHg to a completely normalized 21 mmHg at 3 months. Concurrently, as pulmonary arterial pressure normalized and pulmonary venous return to the left atrium increased, left atrial pressure surpassed right atrial pressure (LAP > RAP). This physiological pressure reversal transformed the interatrial communication from an obligatory, desaturating right-to-left shunt into a

classic, well-tolerated left-to-right shunt across a small secundum ASD, thereby completely eliminating arterial cyanosis and restoring normal systemic oxygen saturation (SpO<sub>2</sub> 98%). Furthermore, with antegrade pulmonary blood flow fully established, the ductus arteriosus underwent spontaneous anatomical constriction and complete fibrous obliteration, entirely bypassing the need for catheter-based or surgical ductal occlusion.<sup>13,28</sup> This documented spontaneous transition confirms that neonates with prenatally diagnosed Ebstein's anomaly who survive the initial transitional window with preserved ventricular function and without circular shunting can achieve stable, unassisted biventricular circulation without emergency neonatal surgery.<sup>7,13,26,32</sup>

In the current surgical era, the decision-making algorithm for neonatal Ebstein's anomaly has been profoundly refined by the consensus recommendations of the American Association for Thoracic Surgery (AATS).<sup>7,8</sup> Historically, symptomatic neonates with severe Ebstein's anomaly faced grim operative prognoses, with initial surgical series reporting perioperative mortality rates approaching 50% to 70%.<sup>1,10</sup> Classical surgical paradigms often defaulted to radical univentricular palliation, such as the Starnes procedure, which entails surgical patch closure of the tricuspid valve orifice, enlargement of the interatrial communication, and construction of a systemic-to-pulmonary arterial shunt (modified Blalock-Taussig shunt), thereby converting the circulation into a functional single-ventricle pathway culminating in the Fontan track.<sup>1,33</sup> However, over the past decade, the worldwide adoption of the cone reconstruction technique pioneered by da Silva and subsequently advanced by Dearani and colleagues has revolutionized outcomes.<sup>3,7,33</sup> The cone procedure achieves complete longitudinal mobilization of all available tricuspid leaflet tissue, surgically detaching anomalous muscular attachments and rotating the mobilized anterior and posterior leaflets 360 degrees to create a circumferential cone of leaflet tissue whose base is re-anchored to the true anatomical atrioventricular junction.<sup>8,33</sup> This provides a near-anatomical, central coaptation zone while plicating the atrialized right ventricle, effectively restoring biventricular circulation.<sup>10,33,34</sup> Nonetheless, neonatal cone reconstruction remains a technically demanding, high-risk intervention. Contemporary consensus emphasizes that surgery during the fragile neonatal window should be strictly reserved for infants with irreversible anatomical right ventricular outflow tract obstruction, refractory low-output circulatory collapse, or profound cyanosis unresponsive to intensive medical therapy.<sup>7,13</sup> In infants who demonstrate antegrade forward flow across the pulmonary valve and achieve adequate systemic oxygenation, the optimal management strategy is aggressive medical bridge therapy—allowing the physiological drop in pulmonary vascular resistance to facilitate natural

right ventricular remodeling and deferring elective surgical repair, if ever required, to late infancy or early childhood when operative mortality drops to under 3% to 5%.<sup>8,10,20</sup> Our clinical experience at RSUD Dr. Saiful Anwar Malang strongly validates this deferred, non-operative physiological bridge paradigm, proving that tertiary centers in developing healthcare systems can achieve optimal survival by avoiding premature neonatal surgical intervention.<sup>7,10</sup>

The concurrent presentation of Ebstein's anomaly with bilateral scrotal hydrocele and suspected hypospadias warrants rigorous developmental consideration. While Ebstein's anomaly is predominantly an isolated defect, non-cardiac congenital anomalies have been reported in 15% to 28% of infants with congenital heart disease in large registry studies, with urogenital anomalies representing a well-recognized subgroup.<sup>35,36</sup> From an embryological standpoint, the critical window for tricuspid valve leaflet delamination (gestational weeks 6 to 10) overlaps temporally with the initiation of external genital differentiation and the formation of the processus vaginalis (gestational weeks 8 to 14).<sup>27,36</sup> The fusion of the urethral folds to form the penile urethra and the descent of the testes into the scrotal sac are highly sensitive to androgenic signaling and local tissue growth factor expression, which are regulated by shared transcription factor networks including BMP, Wnt, and Sonic hedgehog (Shh) pathways.<sup>35,36</sup> A transient environmental or toxic insult during this shared developmental window—such as maternal exposure to endocrine-disrupting combustion byproducts from heavy vehicular traffic—could plausibly perturb both cardiac valve cleavage and urogenital morphogenesis.<sup>25,36,37</sup> Clinically, recognizing that the bilateral hydrocele was a benign, self-limiting accumulation of peritoneal fluid that spontaneously resolved by 3 months post-delivery prevented unnecessary invasive urological intervention during the vulnerable neonatal period.<sup>35,36</sup>

This case also exposes critical health system vulnerabilities and diagnostic screening gaps within the tiered obstetrical network. The delayed identification of a severe congenital heart defect until 34–36 weeks of gestation, coupled with an initial misclassification as DORV, underscores the known limitations of basic obstetric ultrasound screening in non-tertiary facilities.<sup>6,16,38</sup> In accordance with the 2023 updated ISUOG practice guidelines, comprehensive fetal cardiac screening must extend beyond the static four-chamber view to routinely incorporate the five essential diagnostic views: the abdominal situs, the four-chamber view, the left ventricular outflow tract, the right ventricular outflow tract, and the three-vessel and trachea (3VT) view, complemented by color Doppler interrogation.<sup>6</sup> In our patient, marked right atrial enlargement caused significant leftward displacement of the cardiac apex and altered the spatial orientation of the interventricular septum, creating an oblique viewing angle on routine axial sweeps that

mimicked great artery malalignment and gave the false impression of both great vessels originating from the right ventricle (DORV).<sup>6,9</sup> It was only upon multidisciplinary re-evaluation at our tertiary academic institution that specialized fetal echocardiography correctly identified the apical displacement of the tricuspid valve, the atrialized right ventricle, and the normal spatial origin of the pulmonary trunk from the functional right ventricle.<sup>6,9,39</sup> Targeted neonatal echocardiography further provided continuous hemodynamic surveillance, guiding precision fluid administration and pharmacotherapy in the intensive care unit.<sup>40,41</sup> This experience strongly advocates for the widespread implementation of structured fetal cardiac screening protocols and the establishment of multidisciplinary congenital heart disease care networks connecting secondary district hospitals with tertiary academic centers in regional Indonesia, ensuring that fetuses with suspected cardiac anomalies are promptly transferred before labor commences.<sup>42-45</sup>

Finally, the methodological strengths of this investigation include its prospective, longitudinal tracking across four well-defined clinical epochs, the integration of objective multi-parameter prognostic scoring systems (Celermajer Index, SAS, CVPS, Carpentier), and the direct correlation of serial echocardiographic Doppler hemodynamics with clinical outcomes. Limitations include the inherent constraints of a single-case observational design, which precludes statistical generalization across diverse anatomical variants of Ebstein's anomaly. Furthermore, advanced molecular genetic testing (such as chromosomal microarray or whole-exome sequencing to evaluate MYH7, NKX2-5, or GATA4 mutations) was not accessible within the public healthcare framework (BPJS Kesehatan) at our institution. Nevertheless, the granular translational insights derived from this case provide a powerful clinical and biophysical roadmap demonstrating that tertiary multidisciplinary coordination, judicious medical management, and patient non-operative surveillance can achieve exceptional outcomes in prenatally diagnosed Ebstein's anomaly.

## 5. Conclusions

Prenatal identification of Ebstein's anomaly remains a complex clinical challenge that demands heightened diagnostic vigilance during routine obstetric ultrasound screening. When fetal cardiomegaly or four-chamber asymmetry is encountered, clinicians must look beyond conotruncal outflow tract anomalies and systematically evaluate the tricuspid valve insertion plane and ventricular morphology using color Doppler echocardiography in accordance with updated ISUOG guidelines. The favorable clinical course documented in this study underscores that severe neonatal Ebstein's anomaly—even when presenting with elevated pulmonary vascular resistance and massive tricuspid

regurgitation at birth—does not inevitably mandate emergency surgical intervention or carry a fatal prognosis. In the absence of a circular shunt and with preserved functional right ventricular myocardium, the physiological post-delivery decline in pulmonary vascular resistance facilitates dramatic, spontaneous hemodynamic remodeling, characterized by marked tricuspid gradient reduction, spontaneous ductal closure, and interatrial shunt reversal.

Optimal management requires structured multidisciplinary collaboration linking maternal-fetal medicine, pediatric cardiology, neonatology, and obstetric anesthesiology within a specialized tertiary center, ensuring planned term delivery, precision medical stabilization, and longitudinal developmental surveillance. Furthermore, this study demonstrates the vital role of multidisciplinary obstetric-pediatric networks in developing healthcare regions. By bridging secondary community surveillance with tertiary subspecialty care, timely delivery planning and individualized transitional stabilization can be safely delivered. Continued longitudinal surveillance remains imperative through childhood to monitor right ventricular remodeling, rule out delayed supraventricular tachyarrhythmias, and coordinate specialized pediatric urological follow-up.

## Declarations

### *Ethics Approval and Consent to Participate*

This study was conducted in full compliance with the ethical guidelines of the Declaration of Helsinki (2013 Fortaleza revision) and was formally approved by the CMHC Research Center Ethics Committee, Indonesia, under Protocol Approval Number 2026/7482, in coordination with the institutional review board of Dr. Saiful Anwar General Hospital, Malang, East Java, Indonesia.

### *Consent for Publication*

Written informed consent was obtained from the patient and her legal spouse for publication of this observational analytic case study, including all associated anonymized clinical data, electronic cardiotocographic records, fetomaternal ultrasound images, and neonatal echocardiographic findings.

### *Availability of Data and Materials*

All pertinent clinical data, echocardiographic parameters, and analytical frameworks supporting the findings of this study are fully contained within this published article and its accompanying tables and figures.

### *Competing Interests*

The authors declare that they have no competing financial, professional, or personal interests that could influence or appear to influence the work reported in this paper.

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## Authors' Contributions

Conceptualization: NP, QPA, and DK. Investigation, including fetomaternal ultrasound and pediatric echocardiographic evaluation: NP, QPA, and DK. Methodology, designing the multidisciplinary care protocol: NP, QPA, and DK. Resources and clinical management, including obstetrical surgical execution: BR and IWAI. Writing – original draft, data curation, and validation: QPA. Writing – review and editing: BR, IWAI, NP, and DK. All authors read and approved the final manuscript and agree to be accountable for all aspects of the work.

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## Generative AI disclosure

The authors declare that no generative artificial intelligence tools were used in the design, analysis, or writing of this manuscript.

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