Tailored Strategy to Match Anatomy and Physiology with Intervention Can Improve Outcomes of Symptomatic Neonates with Ebstein Anomaly and Tricuspid Valve Dysplasia

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Tailored strategy to match anatomy and physiology with intervention can improve outcomes of symptomatic neonates with Ebstein anomaly (EA)

**Methods**
- Neonatal presentation of Ebstein anomaly (EA) represents the most severe form of this condition.
- Operative mortality remains high and management decisions challenging.

**Results**
- Most common indication for intervention included cyanosis and heart failure 8/8, end-organ dysfunction 8/8, and maldistribution of cardiac output in 6/8. Stabilizing intervention was performed in 3/8. Operative mortality was 1/8.

**Implications**
- At median follow up of 130 months (5-146), there are no late deaths, and all survivors remain in functional class I and free of valvular reintervention.

Management of EA based on anatomy, physiology and preoperative stabilization can help reduce morbidity and mortality in newborns with EA.
Tailored Strategy to Match Anatomy and Physiology with Intervention Can Improve Outcomes of Symptomatic Neonates with Ebstein Anomaly and Tricuspid Valve Dysplasia

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Glossary of abbreviations:

BVR biventricular repair
EA ebstein anomaly
ICU intensive care unit
Kgs kilograms
MPA main pulmonary artery
PA pulmonary artery
RVOT right ventricular outflow

Central Picture legend
Ebstein anomaly of the tricuspid valve in a neonate

Central message:
Tailored strategy to match anatomy and physiology with type and timing of intervention can improve outcomes of symptomatic neonates with Ebstein anomaly

Perspective statement:
Surgical intervention for symptomatic neonates with Ebstein anomaly carries a significant operative risk based on the challenging physiology, technical demands as well as the limited experience with this patient population. A management strategy-based on anatomic features, physiology, preoperative stabilization, and recognition of the deleterious impact of RVOT obstruction may lead to increased survival

Abstract
Objective: Neonatal presentation of Ebstein anomaly represents the most severe form of this condition. Despite significant advances, operative mortality remains high and management decisions represent a formidable challenge. We utilized a strategy aimed to match anatomy and physiology with type and time of intervention to increase survival.

Methods: Review of all patients with fetal or neonatal diagnosis of EA managed at a single
Results: Among 18 patients with EA, 8 underwent neonatal intervention. Most common indication included cyanosis and heart failure (8/8), end-organ dysfunction (6/8) and maldistribution of cardiac output (6/8). Only 2/8 had antegrade pulmonary blood flow. Associated conditions included pulmonary regurgitation in 4/8, atrial tachyarrhythmia in 4 and a VSD in 3. Three patients underwent initial stabilization with MPA occlusion including bilateral PA banding in two. Five patients underwent biventricular repair with conversion to RV exclusion in 2 cases. Three others underwent Starnes procedure as initially planned. Median age at surgery was 10 days (1-30) and median weight 2.6 Kgs (1.9-4.0). Median duration of mechanical ventilation and ICU stay were 9 days (5-34) and 30 days (15-100) respectively. Operative mortality was 1/8. At median follow up of 130 months (5-146), there are no late deaths, and all survivors remain in functional class I and free of valvular reintervention.

Conclusions: Symptomatic neonates with EA can be effectively managed with good outcomes. Preoperative stabilization and choice of management pathway based on anatomy and physiology can help reduce morbidity and mortality.

Word count: 246

Keywords: Ebstein, neonatal, tricuspid valve, surgery
Neonatal presentation of Ebstein anomaly represents the most severe clinical form of this condition characterized by significant impairment in the function of the tricuspid valve, massive cardiomegaly, decreased effective antegrade pulmonary blood flow, right heart failure, metabolic acidosis, as well as end-organ dysfunction. (1,2) Although a significant number of patients can be managed with supportive measures to promote pulmonary blood flow and attain appropriate oxygen delivery during the early period until pulmonary vascular resistance drops, others require an intervention to modify the pathophysiology during this critical period. (3-5) Despite significant advances in surgical technique and perioperative care, overall operative mortality for those individuals who necessitate an intervention remains very high and the decision regarding the optimal management strategy in consideration for a biventricular or a single ventricle management represents a significant challenge. (6-9) Effective surgical techniques to repair the tricuspid valve, (6,10,11) or to exclude the right ventricle (12,13) have been described, however, the risk remains considerable with either approach.

As seen with other difficult choices in the management of newborns with CHD, (14,15) pushing a biventricular strategy in those patients with right ventricular outflow obstruction has met with disappointing results (16,17) and raises the question of selection criteria or the need for modifications with this approach.

Moreover, in contradistinction to other clinical conundrums, the management of these critically ill newborns has remained problematic in no small measure, due to the low volume seen at individual centers. Therefore, it has taken a long time to solve essential questions regarding optimal management pathway, timing of intervention and other technical aspects. (8) We performed a review of our experience based on a strategy that considers anatomic (tricuspid valve morphology and pulmonary atresia) as well as functional variables (antegrade pulmonary blood flow,
pulmonary regurgitation, right ventricular pressure, direction of flow across the PDA) to inform the management and improve the outcomes of this complex group of patients.

Methods:

Between 2007 and 2020, 18 consecutive patients with a fetal or neonatal diagnosis of Ebstein anomaly were managed at our center. All diagnoses were made by echocardiogram utilizing standard criteria, and all fetal and post-natal studies were reviewed by two senior cardiologists (GB, MB). Diagnosis of Ebstein anomaly was based on the apical displacement of the septal and/or posterior leaflet of the tricuspid valve with obvious atrialization of the right ventricle. The presence of additional cardiac defects, namely atrial and ventricular septal defects, pulmonary stenosis or pulmonary atresia, patent ductus arteriosus, left ventricular hypoplasia as well as the presence of antegrade flow across the native pulmonary valve was documented. The diagnosis of anatomic pulmonary atresia was made in the presence of a muscular or membranous plate on 2-dimension images with complete absence of antegrade flow or regurgitation through this area. Functional pulmonary atresia was described as the presence of well-formed pulmonary valve leaflets which did not open and therefore did not exhibit any antegrade pulmonary blood flow or the presence of a normal appearing pulmonary valve with a trace of insufficiency but no antegrade flow.

Patients with corrected transposition of the great arteries and other cardiac anomalies associated with Ebstenoid malformation of the tricuspid valve were excluded. Operative mortality was defined according to the STS Congenital Heart Surgery Database criteria. Primary outcome
measures included 1) operative and late mortality 2) need for reintervention 3) perioperative ECMO support and 4) need for permanent pacemaker.

Follow up was obtained by direct contact with the primary cardiologist as of July 2020. The study was approved by the institutional IRB (Study # 711848-11 4/06/2015) and need for consent was waived.

Results:

Of the 18 patients in this cohort, 16 patients presented with moderate to severe clinical signs. Eight of these were medically managed, allowing eventual closure of the patent ductus arteriosus and progression of antegrade pulmonary blood flow, while exhibiting oxygen saturations above 80%. These patients were subsequently discharged, and all achieved a biventricular circulation. The remaining 8 patients received an operative intervention, which consisted of right ventricular exclusion with a fenestrated patch and a systemic to pulmonary artery shunt (modified Starnes) or a biventricular repair. (Figure 1)

The most common indications for intervention were progressive and/or severe hypoxemia, defined as a PaO2 of less than 30 mmHg while on mechanical ventilation, receiving supplemental oxygen and nitric oxide. Additional criteria for intervention included persistent ductal dependent pulmonary blood flow (beyond a week) as well as heart failure characterized by the presence of pleural effusion, ascites and/or anasarca, maldistribution of cardiac output, persistent lactate elevation and end-organ dysfunction. Nearly half of the patients exhibited new onset of arrhythmias, most commonly supraventricular tachycardia.

All patients met more than two criteria for intervention.
Characteristics of the cohort are shown in Table 1. These characteristics are similar to the previously published national benchmark study, including age and size at first intervention. However, in our cohort the majority had a prenatal diagnosis, and a higher proportion of genetic abnormalities was observed, including a cri-du chat, trisomy X and other chromosomal deletions. Associated anatomic diagnoses were common, including an atrial septal communication in all patients, a ventricular septal defect in nearly half and a patent ductus arteriosus in all but one patient. Anatomic pulmonary atresia was documented in one patient, while only two patients had evidence of antegrade pulmonary blood flow on echocardiogram leaving the diagnosis of functional pulmonary atresia for the remaining 5 patients. Looking at traditional risk criteria, all patients exhibited a cardiothoracic ratio greater than 0.8 on chest x-ray, and a GOS score greater than 1.1, which in association with cyanosis, placed all patients in a high-risk category with near 100% mortality. Regarding the preoperative patient condition (Table 2), a large proportion of patients received preoperative mechanical ventilation and a prostaglandin infusion. A significant number also presented evidence of end-organ dysfunction, most commonly renal, which was evident by persistent and/or progressive creatinine elevation associated with low urine output. As shown in Figure 2, three patients underwent an initial stabilizing intervention, within the first 48 hours of life, to optimize the hemodynamics in preparation for a more definitive intervention with cardiopulmonary bypass. These patients had a median lactate of 6.7, and as high as 12 mmol/dl, due to circulatory embarrassment largely associated with a circular shunt in the presence of significant tricuspid regurgitation, pulmonary valve regurgitation and a patent ductus arteriosus. The stabilizing interventions included clip occlusion of the pulmonary valve in three patients, one
of whom also received a bilateral pulmonary artery banding. These interventions were aimed at elimination of the circular shunt and control of the pulmonary blood flow to facilitate the overall balance of distribution of cardiac output, which commonly led to normalization of lactates and a prompt recovery of end-organ dysfunction. The median preoperative lactate at the time of the main intervention was 1.5 mg/dl.

**Surgical Intervention:**
Choice of surgical strategy was based on a combination of anatomic and physiologic variables. In broad terms, a biventricular repair was planned in the presence of a functional tripartite RV with a patent outflow and an estimated RV pressure greater than 35 mm of Hg. Patients left the operating room with a biventricular repair unless the residual tricuspid regurgitation was greater than moderate, the tricuspid valve repair was not deemed durable and/or there was inadequate effective pulmonary blood flow associated with severe hypoxemia. In contradiction, patients with a functional unipartite RV, with poor function and muscular pulmonary atresia were planned for RV exclusion. (Figure 3) Five patients were scheduled to have a biventricular repair while right ventricular exclusion was planned in three. Among those planned for a biventricular repair, one patient underwent a conventional biventricular repair creating a bi-leaflet tricuspid valve with commissural plication of as well as a Sebening stitch. (6) The remaining individuals underwent a Cone reconstruction (Video 1) with circumferential leaflet coverage and posterior annular plication, based on the original description of the technique. (10) However, this was modified to avoid the plication of the atrialized portion of the right ventricle as well as a circumferential annuloplasty, to permit future growth of the tricuspid valve. (11) Two of these patients had an intraoperative conversion from BVR to a right ventricular exclusion due to inadequate tissue to
pursue a Cone repair or the reconstruction was deemed non-satisfactory due to moderate-to-severe residual regurgitation. Therefore, three patients left the operating room with a biventricular repair while five received a single-ventricle palliation which consisted of a right ventricular exclusion with a systemic to pulmonary artery shunt. (Figure 4) The source of pulmonary blood flow was a 3.5 mm right modified BT shunt in four patients, while a central shunt was used in a patient with a chromosomal anomaly, severe branch pulmonary artery hypoplasia and left diaphragmatic eventration (Table 3)

The median aortic cross-clamp was 51 minutes, and the median cardiopulmonary bypass duration of 110 minutes. Analysis of the perfusion conduct during the operative intervention revealed that while the duration of cardiopulmonary bypass was not significantly different for each strategy, although patients undergoing biventricular repair had a median period of myocardial ischemia about 20 minutes longer compared to those undergoing single-ventricle palliation, even if intraoperative conversion had occurred. This was likely a reflection of the fact that the decision to pursue conversion was made relatively early during the operation and while attempting to minimize the period of myocardial ischemia in these vulnerable newborns. A delayed sternal closure was performed in nearly all (6/8) patients.

Outcomes

As shown in Table 4, there was a single death out of 8 patients. This was a low-birth-weight neonate with EA and trisomy X, who had low Apgar scores, pulmonary atresia, hypoplastic central pulmonary arteries associated with moderate lung hypoplasia and a diaphragmatic eventration, who in the early phase of this experience underwent right ventricular exclusion and placement of a 4 mm central shunt due to anticipated difficulty in providing enough pulmonary blood flow.
Despite appropriate hemodynamics and gas exchange in the first 24 hours, the patient exhibited acute decompensation secondary to ectopic atrial tachycardia which required e-CPR and subsequently exhibited maldistribution of cardiac output, acute kidney injury and subsequently expired on postoperative day #37.

Two patients received postoperative mechanical circulatory support. One of these just was described. A second patient with a birthweight of 2.3 kg and cri du chat underwent initial stabilization by clip occlusion of the pulmonary valve due to maldistribution of cardiac output associated with severe TR and a circular shunt, followed by right ventricular exclusion with a 3.5 mm right modified BT shunt. Early in the postoperative period presented with acute hemodynamic decompensation secondary to supraventricular tachyarrhythmia for which he received emergency ECMO cannulation. Following 48 hours of support, the patient was successfully weaned, then discharged and eventually underwent Fontan completion.

Unplanned cardiac reoperations occurred in two patients. One patient who received ECMO support underwent right ventricular reduction due to progressive RV dilatation following Starnes procedure, while another required evacuation of cardiac tamponade in the immediate postoperative period. Two patients exhibited postoperative worsening of the preoperative acute kidney injury (creatinine >2.0 from baseline and/or urine output < 0.5 ml/kg/hr for > 12 hrs), but did not require replacement therapy. There were no shunt related reoperations or reinterventions and no permanent pacemaker implantation.

At median follow up of 130 months (5-146), there are no late deaths, and all survivors remain in functional class I and free of valvular reintervention. (Figure 5).
Neonates with EA exhibit a complex physiology which is multifactorial, including the presence of severe tricuspid valve regurgitation, and physiological elevation pulmonary vascular resistance of the newborn. This is aggravated by abnormal lung development and pulmonary hypoplasia, with reduced generation of alveoli and alveolar simplification. (19) Moreover, in cases of EA, a myopathy of the right ventricular muscle can add to the inability to achieve effective antegrade pulmonary blood flow. Ineffective filling of the left ventricle and the frequent association of supraventricular arrhythmias can further aggravate a very precarious circulatory balance, which in the presence of pulmonary regurgitation and a circular shunt could become unmanageable. Not surprisingly, the reported mortality rate for these symptomatic neonates remains very high. (1,2,18,20). In addition, the severity of the circulatory compromise can become evident in utero, with signs of heart failure, hydrops, and even fetal demise. These prenatal data convey a grim prognosis and has frequently led to a high rate of termination, further compromising the outlook of patients with this condition. (5,21)

While the ideal management strategy for these symptomatic neonates might be supportive allowing time for the physiologic decrease in pulmonary vascular resistance, this may not be attainable in some patients. Particularly in those who present with anatomic obstruction of the RVOT, those with right ventricle dysfunction or in those who exhibit circulatory compromise associated with arrhythmias or the presence of a circular shunt. In this cohort of 18 neonates, roughly half of those with clinical signs received medical management and achieved a biventricular circulation. This compares favorably with the PHIS cohort of over four hundred patients of whom nearly two thirds were managed medically with a 22% early mortality (20)
Although this could be attributed to an era effect, this difference in mortality may raise questions about the selection criteria to pursue medical management and perhaps the possibility that some of these newborns may have benefited from a timely surgical intervention.

The other half of our cohort underwent neonatal intervention, which consisted of single ventricle palliation or biventricular repair with a 2:1 ratio. The anatomic characteristic of the tricuspid valve, contractility of the right ventricle (estimated right ventricular pressure > 35 mm Hg in the absence of a ventricular septal defect), patency of the right ventricular outflow tract, direction of flow across the patent ductus as well as patient’s condition at the time of intervention were important considerations when planning for the type and timing of the surgical intervention. One strategy is to achieve a biventricular circulation based on the repair of the tricuspid valve (6) while alternatively those newborns with less favorable anatomy usually undergo a right ventricular exclusion procedure or insertion of a systemic to pulmonary artery shunt alone if the predominant issue is hypoxia in the absence of heart failure. Using the STS Congenital Heart Surgery Database as a reference allowed us to ascertain the overall applicability of our observations against the benchmark experience with surgical management of EA in neonates. (18) Although a higher incidence of prenatal diagnosis and chromosomal anomalies were evident in our cohort, their impact remains unclear. While prenatal diagnosis can lead to a higher level of preparedness, particularly for those with ductal dependent pulmonary circulation, it also has led to an increased frequency of interruption of pregnancy, (5,21) therefore the overall benefit of prenatal diagnosis on patient survival remains debatable. Additionally, an increased incidence of chromosomal anomalies has conferred an additional level of risk when it comes to outcomes in these complex newborns, (22) particularly when this is associated with low birth weight. This has been documented by Curzon, who reported that low birth weight newborns with EA undergoing a
systemic to pulmonary artery shunt have seven-fold increase in mortality. (9) Although the median weight in our cohort was 2.7 kgs and as low as 1.9 kgs, weight did not observe a significant impact on outcome. We would urge caution about this observation that perhaps could be explained by the deliberate measures taken to optimize overall patient condition and end-organ function prior to surgery, as well a relatively short period of myocardial ischemia and a conservative management strategy in those newborns with pulmonary atresia.

Unfortunately, one of the biggest challenges we confronted was the high incidence of perioperative arrhythmias, which were particularly destabilizing, and led to e-CPR in two cases. Management of arrhythmias has proven to be particularly difficult in those patients with a circular shunt and/or during the postoperative period, nevertheless no preemptive treatment strategies have been described, nor were utilized at our center to address this issue.

As expected, the overall preoperative condition had significant influence on the overall outcome of these patients. About three-quarters of the patients exhibited inadequate circulation with significant lactate level elevation and important end-organ dysfunction preoperatively. Directed interventions to eliminate a circular shunt (23,24) and/or balance the circulation by controlling pulmonary blood flow proved to be lifesaving and had a profound salutary effect, allowing patients to arrive at the main intervention in an elective fashion after optimization of organ function had occurred. It should be noted that in patients with anatomic pulmonary atresia creation of pulmonary insufficiency by perforation of the pulmonary valve plate would be associated with the possibility of a circular shunt and therefore should be avoided. Although bilateral pulmonary artery banding may seem completely counterintuitive, it has become increasingly apparent that many patients who present with a circular shunt and severe volume overload, may be affected by maldistribution of cardiac output, extreme systemic venous desaturation, and coronary perfusion issues which
ultimately contribute to unremitting shock. It has been our observation that once the circular shunt is eliminated, PaO2 levels can rise above 35 mm Hg and therefore creating the opportunity to balance the pulmonary blood flow and improve coronary and systemic perfusion. This observation has been replicated by Hasegawa and colleagues in a patient with prenatal diagnosis of severe hydrops. (25) and is similar to the initial intervention described in neonates who present in shock with delayed diagnosis of a ductal dependent systemic circulation. (26) Alternatively, ECMO support can provide the necessary stabilization, while avoiding the adverse effects associated with extreme ventilatory measures and suboptimal perfusion, particularly if surgery is anticipated in the immediate future. Success with preoperative circulatory stabilization has been previously reported, including the successful management of a newborn with a significant degree of hydrops at birth. (27,28) As the management of these sick neonates continues to improve, traditional risk criteria to predict outcomes like cardiothoracic ratio on CXR and GOS echocardiographic score has been overcome, making predictions quite difficult. New imaging modalities that hold promise include lung volumetric calculation of the lung parenchyma by MRI as well as evaluation of the pulmonary artery size and direction of flow. (29,30) Over the last 30 years, distinct surgical approaches have been utilized largely based on anatomic variables including the size of the tricuspid valve a, the size of the functional (nonatrialized) right ventricular cavity and the presence of pulmonary atresia. (1,12) It has been clearly demonstrated that pulmonary atresia confers a two-fold increase in mortality following BVR (16) and should be considered a significant prognostic factor when it comes to deciding the type of intervention. More recently, physiologic inferences by echocardiogram suggest that an estimated RV pressure > 30 mm Hg and antegrade flow across the RVOT and pulmonary valve are associated with better
outcomes following BVR. (13) In our cohort, we utilized these variables along with a careful consideration for preoperative patient condition to plan on a management strategy we believed would be the most reproducible and likely associated with survival.

While the strategy of BVR has been traditionally based on the creation of a double-orifice valve with some partial leaflet attachment and rotation as well as augmentation and annuloplasty, significant improvements were achieved with a better understanding of the plane of coaptation and management of the right ventricular outflow obstruction. Moreover, the introduction of the Cone reconstruction has improved the reproducibility of the tricuspid valve repair even in the newborn, having a favorable impact on outcomes. (10,11,28) Additionally, successful conversion from RV exclusion to biventricular repair has been described, raising the option of a staged approach to mitigate the additional challenge of biventricular repair in the newborn. (31)

In conclusion, while clinical presentation of EA in the neonate is associated with life-threatening physiology, an initial intervention aimed at physiologic stabilization and recovery of end-organ function can have a favorable impact on outcomes mitigating morbidity and overall mortality. The choice of management pathway between right ventricular exclusion versus biventricular intervention should be based on anatomic and physiologic variables rather than on arbitrary anatomic criteria. Risk markers traditionally associated with a poor outcome are no longer valid and new predictors should be identified to better inform decisions and counsel parents about the management options and their likelihood of success in patients with this complex form of congenital heart disease.
References


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Cone procedure and ventricular septal defect closure on a neonate with Ebstein anomaly

Figure 1
Entire cohort and management pathway

Figure 2
Initial interventions to optimize the physiology

MPA, main pulmonary artery; bPAB, bilateral pulmonary artery banding; EA, Ebstein anomaly;
BVR, biventricular repair

Figure 3
Management algorithm

Figure 4
Planned surgical interventions versus final intervention

BVR, biventricular repair; SVR, single ventricle reconstruction

Figure 5
Graphical abstract

Supplemental Table 1
Clinical cohort evolution
<table>
<thead>
<tr>
<th>Study Cohort</th>
<th>Median (range)</th>
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</thead>
<tbody>
<tr>
<td>Age (days)</td>
<td>10.0 (1-30)</td>
</tr>
<tr>
<td>Male (%)</td>
<td>54.5</td>
</tr>
<tr>
<td>Weight (kg)</td>
<td>2.6 (2.0-4)</td>
</tr>
<tr>
<td>Gestational age (weeks)</td>
<td>37.0 (35-40)</td>
</tr>
<tr>
<td>Prenatal diagnosis (%)</td>
<td>87.5 (7/8)</td>
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<tr>
<td>Genetic abnormality (%)</td>
<td>37.5 (3/8)</td>
</tr>
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</table>
### TABLE 2. Preoperative Variables

<table>
<thead>
<tr>
<th>Study Cohort</th>
<th>Median (range)</th>
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</thead>
<tbody>
<tr>
<td>ECMO support (%)</td>
<td>none (0/8)</td>
</tr>
<tr>
<td>Pre op ventilation (%)</td>
<td>87.5 (7/8)</td>
</tr>
<tr>
<td>PGE1 dependent (%)</td>
<td>62.5 (5/8)</td>
</tr>
<tr>
<td>Cyanosis (%)</td>
<td>100 (8/8)</td>
</tr>
<tr>
<td>Circular shunt (%)</td>
<td>50 (4/8)</td>
</tr>
<tr>
<td>Shock (%)</td>
<td>12.5 (1/8)</td>
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<tr>
<td>Resolved shock (%)</td>
<td>50 (4/8)</td>
</tr>
<tr>
<td>Organ dysfunction (%)</td>
<td>87.5 (7/8)</td>
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<tr>
<td>Does not include hypoxemia</td>
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<tr>
<td>Peak lactate (mmol/dl)</td>
<td>6.0 (3.5-12)</td>
</tr>
<tr>
<td>Stabilizing intervention (%)</td>
<td>37.5 (3/8)</td>
</tr>
<tr>
<td>RV pressure &gt; 35 mmHg (%)</td>
<td>62 (5/8)</td>
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ECMO, extracorporeal membrane oxygenation support; preop, preoperative; PGE1, prostaglandin E1;
TABLE 3. Intraoperative variables

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<thead>
<tr>
<th>Study Cohort</th>
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<tr>
<td>Pre-operative lactate (mmol/dl)</td>
<td>1.8 (2.0-1.0)</td>
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<tr>
<td>Aortic cross clamp (min)</td>
<td>51 (24-105)</td>
</tr>
<tr>
<td>CPB (min)</td>
<td>110 (75-192)</td>
</tr>
<tr>
<td>Shunt size (mm)</td>
<td>3.5 (3.0-4.0)</td>
</tr>
<tr>
<td>Central shunt (%)</td>
<td>12.5 (1/8)</td>
</tr>
<tr>
<td>Open sternum (%)</td>
<td>75 (6/8)</td>
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CPB, cardiopulmonary bypass
### TABLE 4. Surgical outcome measures

<table>
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<tbody>
<tr>
<td>Operative mortality (%)</td>
<td>1/8</td>
</tr>
<tr>
<td>Post op mechanical support (%)</td>
<td>E-cpr x 2 / rhythm related</td>
</tr>
<tr>
<td>Unplanned cardiac reoperation (%)</td>
<td>RV reduction x 1, tamponade x 1</td>
</tr>
<tr>
<td>Reoperation for shunt placement or revision (%)</td>
<td>None</td>
</tr>
<tr>
<td>AKI (%)</td>
<td>25 (2/8)</td>
</tr>
<tr>
<td>Permanent pacemaker (%)</td>
<td>None</td>
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</table>

Post op, postoperative; E-cpr, cardiopulmonary resuscitation with extracorporeal support; TV, tricuspid valve; RV to PA, right ventricle to pulmonary artery; AKI, acute kidney injury.
Initial stabilizing intervention (resuscitation)

- Median age 1.5 days (1-10)
- Median lactate 6.7 mmol/dl (3.6-12)
- Organ dysfunction 4/8
Surgical Procedures

Planned

1. Ebstein repair
2. Cone reconstruction
3. Starnes procedure

Achieved

1. BVR
2. BVR
5. SV
Tailored strategy to match anatomy and physiology with intervention can improve outcomes of symptomatic neonates with Ebstein anomaly (EA)

<table>
<thead>
<tr>
<th>Methods</th>
<th>Results</th>
<th>Implications</th>
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| - Neonatal presentation of Ebstein anomaly (EA) represents the most severe form of this condition.  
  - Operative mortality remains high and management decisions challenging. | Most common indication for intervention included cyanosis and heart failure 8/8, end-organ dysfunction 8/8 and maldistribution of cardiac output in 6/8. Stabilizing intervention was performed in 3/8. Operative mortality was 1/8. | At median follow up of 130 months (5-146), there are no late deaths, and all survivors remain in functional class I and free of valvular reintervention. |

Management of EA based on anatomy, physiology and preoperative stabilization can help reduce morbidity and mortality in newborns with EA.
Tailored Strategy to Match Anatomy and Physiology with Intervention Can Improve Outcomes of Symptomatic Neonates with Ebstein Anomaly and Tricuspid Valve Dysplasia

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