Commentary: Neonates with Ebstein anomaly: a paradigm strategy of Starnes procedure to be followed by bi-ventricular repair

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Central Message: Critically sick neonates with Ebstein anomaly and circular shunt can be stabilized with Starnes procedure yet considered for subsequent bi-ventricular repair.

Central Picture legend: Modified Starnes operation with preservation of pulmonary and tricuspid valves.

Abbreviations
ASD – atrial septal defect
RV – right ventricle
PA – pulmonary artery
ECMO – extracorporeal membrane oxygenation
TR – tricuspid regurgitation

Critically sick neonates with Ebstein anomaly and circular shunt (Figure 1A), require urgent surgical treatment, and Starnes operation delivers predictably good stabilization to permit subsequent surgical management (1-4). It becomes increasingly clear that surgical management after the initial life-saving Starnes operation should aim at bi-ventricular repair, if feasible. Herein a group from Columbia University recommended that all patients who underwent the Starnes procedure should be systematically evaluated for subsequent bi-ventricular conversion (5).

A few comments appear appropriate. While successful neonatal bi-ventricular repair can be accomplished in selected patients by a highly experienced team (6-10), Starnes procedure provides stabilization for the majority of neonates. The Starnes procedure is an important and reproducible first step to stabilize the critically sick neonate, particularly at institutions with limited experience in management of neonates with Ebstein anomaly. Most importantly, Starnes operation does not preclude subsequent bi-ventricular repair, or 1.5 ventricle repair if a bidirectional cavo-
pulmonary shunt (BCPS) has already been performed (2,7, 10-14). In fact, we believe that every patient that had a Starnes operation should be evaluated for subsequent bi-ventricular repair. This does not imply that every patient must have bi-ventricular repair, but at the current time, no patient should be automatically committed to uni-ventricular pathway, without proper evaluation and consideration of bi-ventricular repair first. This staged management raises important questions. Is there anything that can be done during the Starnes procedure to facilitate subsequent bi-ventricular repair? What are the criteria for selecting the patients suitable for bi-ventricular repair? What is the optimal timing for bi-ventricular or 1.5-ventricle repair?

In as much as we are entering uncharted territory, sharing some thoughts appear to be appropriate. As the first decision on whether to proceed to bi-ventricular repair or BCPS will be done in infancy, it is important to modify the original Starnes operation to achieve optimal preservation of the pulmonary and tricuspid valves as well as to ensure the age-appropriate growth of the right ventricular cavity. Pulmonary valve preservation with intraluminal patching of the main pulmonary artery if pulmonary regurgitation is present (Figure 1B) and also placement of fenestrated polytetrafluoroethylene patch further away from tricuspid valve leaflets may reduce potential scarring between the patch and leaflets and facilitate subsequent bi-ventricular repair (2,13). The patch is sutured above the tricuspid valve annulus along the Todaro ligament (Figure 1C-D) usually leaving the coronary sinus orifice in the right atrium. This strategy allows some involution of the right ventricle that is advantageous for a univentricular pathway. However, controversy remains regarding the location of the coronary sinus orifice relative to the patch since leaving the coronary sinus on the right ventricular side (Figure 1E) allows some volume loading of the right ventricle that may facilitate sufficient growth of the right ventricle to allow a bi-ventricular repair. Ideally,
bi-ventricular repair should be performed in during infancy in suitable patients. However, if the right ventricle appears too small, as the infant begins to overgrow the systemic-to-pulmonary shunt (Figure 2A), a fenestration can be added to the intraluminal patch of the main pulmonary artery at the time of BCPS (Figure 2B) to facilitated volume loading and growth of the right ventricle that can be rehabilitated toward 1.5-ventricle (Figure 2C) or 2-ventricle circulation (Figure 2D). The exact modes of assessment, algorithms and timing for bi-ventricular repair following Starnes operation are yet to be elucidated. Yet, conceptually, it appears timely to emphasize that every effort should be made to potentially allow bi-ventricular repair in children after the initial Starnes operation.

References


modifications to facilitate subsequent biventricular repair of Ebstein anomaly.


**Figure legends**

**Figure 1.** Modifications to Starnes operation to facilitate subsequent bi-ventricular repair.

A. Circular shunt in a neonate with Ebstein anomaly.

B. Intraluminal banding of the main pulmonary artery.

C. Closure of the right ventricular inflow tract with a fenestrated patched placed into the supravalvular position and sutured to the tendon of Todaro to avoid damaging conduction system or tricuspid valve leaflets.


**Figure 2.** Placement of fenestration in the intraluminal patch of the main pulmonary artery at the time of bi-directional cavopulmonary shunt. This strategy may allow some volume load of the small right ventricle that can be rehabilitated for subsequent 1.5- or 2-ventricle repair. Modified Starnes operation with intraluminal patch closure of the main pulmonary artery is performed (A). Fenestration to the intraluminal patch closing the main pulmonary artery can be added at the stage of bidirectional cavo-pulmonary shunt to allow some forward flow in the pulmonary artery and loading of the right ventricle (B). The intraluminal patch is removed from the main pulmonary artery, atrial
septal defect is closed, and tricuspid valve is reconstructed to achieve either one and one-half ventricle repair (C) or bi-ventricular repair (D).

RA – right atrium, RV - right ventricle; SVC – superior vena cava.

Central Picture. Modified Starnes operation with preservation of pulmonary and tricuspid valves.
A. RA resected

B. Ventricular rehabilitation

C. SVC

D. Biventricular repair

Biventricular repair